Children's Heart Unit for Wales
Uned Calon Plant I Gymru

Clinical Guidelines

From March 2023

These Guidelines have been reviewed by the Paediatric Cardiology consultant team and Executive members of the Welsh Paediatric Cardiovascular Network – they are reviewed on a regular basis and are intended to be used to assist in the management of children with heart problems or suspected heart problems.
The Acute Child Health Directorate at Cardiff and Vale University Local Health Board has indicated its approval for these guidelines to be used in combination with clinical judgement and experience.

GIG CYMRU NHS WALES
Bwrdd Iechyd Prifysgol Caerdydd a'r Fro
Cardiff and Vale University Health Board

Contents

SECTION 1 – ADMINISTRATION

1.1 UHW Telephone Numbers

1.1.1 Consultant Medical Staff and Speciality Doctor

Clinician	Secretary	Office	Radiopager	Mobile
Dr Chris Gillett	41239	41682	02921 287 238 (Skype line)	Via UHW Switchboard
Dr Nadia Hajiani	44606	41742	-	
Dr Victor Ofoe	44743	43869	07623 905 928	
Dr Alan Pateman	44606	42908	07623 906 018	
Prof Orhan Uzun	44743	44745	07623 906 121	
Dr Dirk G Wilson	44749	45156	07623 905 734	
Dr Amos Wong	47750	46985	Bleep 6343	

- Preferred contact is via mobile phones or radiopager.
- If all efforts to contact the on-call consultant are unsuccessful, contact one of the other consultants; in a dire emergency, speak to the on-call Bristol consultant for advice.

1.1.2 Adult Congenital Heart Disease Staff

The ACHD consultant team is comprised of Dr Helen Wallis, Dr Simon MacDonald, Dr Nav Masani and Dr Dirk Wilson. The ACHD coordinator is Mrs Elizabeth Corris – extension 43892.

1.1.3 Secretarial Staff

The clinical secretaries are located in Cardigan House.

Name	Secretary to	Extension
Amanda Doyle	Dr Dirk Wilson	44749
Caroline Keogh	Prof Orhan Uzun Dr Victor Ofoe	44743
Julie Taylor	Dr Chris Gillett	41239
Emma Wakefield	Dr Alan Pateman Dr Nadia Hajiani	44606
Lynn Page	Support secretary, including Clinical Psychology	47750
Hannah Loach	Support secretary	-

1.1.4 Junior Medical Staff/Trainees

Title	Bleep	Extension
Cardiac Registrar	5391	44761
Paediatric Registrar	5391	44761
SHO/ST2-3	5391	44761

1.1.5 Psychology

We are supported by a team of psychology staff. If you think a patient may benefit from psychology input, discuss it with the named specialist nurse and consultant. There is a weekly referral meeting where the case can be discussed. There is a referral form on the Shared Directory: S:\PaedCard\SHARED\PSYCHOLOGY\REFERRAL FORM

1.1.6 Technical and Support Staff

Staff	Bleep/Radiopager	Extension
Cardiac Physiology (echo) Claire Gemmell and Leann Gibbs	Contact via WhatsApp	43920
Cardiac Dieticians	07623 905 696	46995

1.1.7 Specialist Cardiac Nurses

There is a team of specialist cardiac nurses, some of whom are full-time and some of whom are part-time. Some provide support for a specialised area, while others provide support for specific geographical areas.

Area / Speciality	Nurse	Extn	Mobile
Aneurin Bevan	Karina Parsons-Simmonds	45524	07966 461 421
Cardiff and Vale	Claire Logan (Team Leader) [locality cross-cover by Emma Benger]	43920	07811 197 136
Cwm Taf Morgannwg	Emma Benger	47021	07890 026 021
Hywel Dda	Wendy Williams and Jenni Stirling	44753	07813 922 441
Swansea Bay			07813 922 441
Fetal	Alison Pearce	41746	07814 773 446
Transition (all areas, age 13+)	Karina Howell Laura McCarthy	48046	07980 635 177

Generic email: paediatriccardiac.cns@wales.nhs.uk

Roles and Responsibilities

- Nurse-led clinic (see Clinics, Section 3.2)
- Primary contact in INR service (section 6.1)
- Link between clinicians and parents
- Link between Cardiff and Bristol – participation in weekly Planning Meeting
- Parent education and advice
- Oversight of Home Monitoring Programme

The cardiac liaison nurses should be contacted about:

- Newly diagnosed children
- Cardiac admissions to the ward
- Cardiac catheter or operation preparation
- Any decision to transfer a patient (so they can keep track of transfers out)
- Distressed or anxious children or parents
- Ward discharges
- Patients experiencing difficulty with transition care
- Stethoscope training for heart rhythm assessment
- Discussions about psychology support
- Discussions about palliative management
- Oversight of 6-minute walk test

The adult congenital cardiac nurse specialist team is based in Cardiology Outpatients. Contact details are:

Landline	029 2184 4580
Mobile	07966 382 995
Email	achdnurse.cav@wales.nhs.uk

Roles and Responsibilities

- Supporting ACHD clinics (UHW and outreach)
- Information and advice for ACHD patients / families
- Link between Cardiff and Bristol ACHD services
- Support for inpatients
- Pre- and post-operative support
- Support for patients with learning difficulties, including mental capacity and best interest assessment
- Discussions about psychology support
- Discussions about palliative management
- Provision of nurse-led clinics
- Oversight of 6-minute walk test

1.1.8 Useful Contact Numbers

Use the Induction hospital app (available free of charge on app stores)
Paediatric Clinical Areas

Ward Name	Telephone Numbers
Children's Assessment Unit (Seahorse)	45441/45426
Children's Emergency Department	48018
Clinical Investigation Unit (Seahorse)	43765
Island Ward (General Paeds)	43359/45330/48859
Flow manager	Bleep 6434
Jungle Ward (General Paeds)	43274/43276
Owl / Gwdihw Ward (Surgery)	43277/42650/47348
Pelican Ward (Cardiac/Renal)	44755/45375
Paediatric Critical Care Unit	44751 (HDU) 43282/44622/45323
Rainbow Ward (Oncology)	48801/48832
Rocket Outpatients (Oncology)	48805
Starfish Outpatients	43364/44164
Theatres (Paediatric)	47457 / 47337

Department/Individual	Extension	Bleep	Other Information
Adult Cardiology B1 Ward (inpatient cardiology) Cardiac ICU Cardiac Day Case Unit Catheter Laboratory Coronary Care Unit	 43382/44603 43265 44414 43329/44607 42110		

Ambulance Control	43777 0300 123 9234		Booked routine Emergency
Anaesthetic Department	43107		
Biochemistry Acute Biochem Lab Main Office Lab Reception Duty Biochemist	 42637 42909 42805 48334	 5452	 Call Lab for urgent processing
Blood Bank	42157/42158	5268	
CARDIAC ARREST	2222		
CHANTS Neonatal Transport	Call 42680 and ask which team is on call		1:3 Rota Cardiff 02920 744719 Newport 01633 234844 Swansea 01792 285278/5403
Children's Assessment Unit	45441		
Coagulation	46477	5270	Coag Reg bleep 5886
Coroner's Office	01443 281 100		Outside line required
Delivery Suite	42686		
Dental Clinic (paeds)	42458		
Drug Information	44328		
ECG (Main Dept, Pacing)	45489		
ECG (Inpatient requests)	43198		Suite 11 OPD (Holter also)
EEG/Neurophysiology	43194		
Exercise/Tilt Test	43465		
Fetal Medicine	42279/43341		
Flow manager `		6434	
Genetics Clinical Lab Reception	 44036 44023 42577		
Haematology Emergency o-o-h Lab Paed SpR	 5269 46477	 6517	 Bleep for out-of-hours samples
Hand Over Room General	48930		
Hand Over Room Specialty	48820		
Histology	42710/42714		
Holter (24-hour tape)	46695		
IT help desk	44000		
Main Theatres Reception	42993		
Media Resources	43305/43307		
Microbiology Lab	42044		
MRI (Paeds)	47338		

Neonatal Unit Reception	42680/42684		
Buttercup (HDU)	46873/46771		
Bluebell (ICU)	41751		
Daisy (Low Dependency) (Nursery)	45911 45910		
OPD (Adult Cardiac)	43266		Adult Congenital Clinic
OPD (Starfish)	43364/44164		
Pacing Clinic	44457/45489		
Pacing Lab	43081		
Paediatric ICU	43282/44622		
Paediatric Theatres Reception	47357		
Pathology Reception	47397		
Personnel (Human Resources)	43887/45300		
Pharmacy	42988		
Porters	42667		
Postnatal Ward	43343		
Public Health	42236		
SALT team	43736		
Sophie Pearson Room	46355		
Switchboard	100		Emergency 2222
Teenage Cancer Trust	46784/46915		Dr's Office 42973
Toxicology	26894		Drug levels
Ultrasound	44834		
WATCH PICU/HDU transport			03000300789
X-ray (Paeds)	43953	5299	Out of hours – call extn 48064

1.2 Welsh Hospitals

District General Hospital	WHTN	External Number
Aberdare General Hospital	01753	01685 872 411
Brecon War Memorial Hospital	01762	01874 622 443
Bronglais General Hospital (Aberystwyth)	01822	01970 623 131
Grange University Hospital (Cwmbran)	-	01633 493 100
Llandough Hospital	01776 (From UHW dial "2" then the 4 digit extn)	029 2071 1711
Morriston Hospital (Swansea)	01789	01792 702 222
Neath Port Talbot Hospital	01885	01639 862 000

Nevill Hall Hospital (Abergavenny)	01835 5800	01873 732 732
Prince Charles Hospital (Merthyr)	01854	01685 721 721
Princess of Wales Hospital (Bridgend)	01855	01656 752 752
Prince Phillip Hospital (Llanelli)	01824	01554 756 567
Royal Glamorgan Hospital	01751	01443 443 443
Royal Gwent Hospital (Newport)	01835 5800	01633 234 234
Saint David's Hospital	01771	029 2053 6666
Singleton Hospital (Swansea)	01883	01792 205 666
West Wales General Hospital (Carmarthen)	01827	01267 235 151
Withybush Hospital (Haverfordwest)	01720	01437 764 545
Wrexham Maelor Hospital	01814	01978 292000
Ysbyty Cwm Rhondda (Llwynypia)	01803	01443 430 022
Ysbyty Glan Clwyd (Rhyl)	01815	01745 583 910
Ysbyty Gwynedd (Bangor)	01746	01248 384 384
Ysbyty Ystrad Fawr (Ystrad Mynach)	01835 5800	01443 802200

Add "100" to the WHTN number for Switchboard Operator or direct-dial if you know the extension

1.3 Bristol Children's Hospital

Useful Bristol Numbers

Switchboard	01179 215 411 or 01179 276 998
Catheter Laboratory (BRHC)	0117 342 8282 / 8456
CNS Team	01179 342 8578
Doctors' Office (Ward 32, BRHC)	0117 342 8196
Echocardiography Laboratory (BRHC)	0117 342 8722 /8181
Paediatric ICU	0117 342 8377 or 8437
Paediatric Cardiac Ward (32, BRHC)	0117 342 8332 / 8679
Paeds OPD	0117 342 8401 or 8402
SCBU/NICU (St Michael's)	0117 342 5275 or 5275
Secretaries Cardiology Cardiac surgical	0117 342 8853 0117 342 8977
BHI Coronary Care Unit (BHI)	0117 342 2278
BHI GUCH Liaison Nurse (BHI)	0117 342 0463

1.4 UK and Ireland Paediatric Cardiac Units

Centre	Switchboard
Alder Hey Children's Hospital (Liverpool)	0151 228 4811
Birmingham Children's Hospital	0121 333 9999
Bristol Royal Hospital for Children	01179 230 000 01179 276 998 01173 428 460
Evelina Children's Hospital (Guys Hospital, London)	0207 188 7188
Freeman Hospital (Newcastle-upon-Tyne)	0191 233 6161
Glenfield Hospital (Leicester)	0300 303 1573
Great Ormond Street Hospital for Children (London)	0207 405 9200
Harefield Hospital (London)	01895 823 737
John Radcliffe Hospital (Oxford)	0300 304 7777
Leeds Congenital Heart Unit	0113 392 5467
Liverpool Heart and Chest Hospital (ACHD service)	0151 600 1616
Our Lady's Hospital for Sick Children (Dublin)	00 353 1409 6100
Royal Belfast Hospital for Sick Children	02890 240 503
Royal Brompton & Harefield NHS Trust (London)	0207 352 8121
Royal Hospital for Sick Children (Edinburgh)	0131 536 0000
Royal Hospital for Sick Children (Yorkhill, Glasgow)	0141 201 0000
Royal Manchester Children's Hospital	0161 701 2179
Southampton Children's Hospital	02380 777 222

1.5 Paediatricians/Neonatologists with Expertise

Local Health Board	**Hospital**	**Doctor**	**Email address**
Aneurin Bevan UHB	Nevill Hall Royal Gwent	Dr Soha El-Behery Dr Sandeep Ashtekar Dr Marion Schmidt Dr Kevin Poon Dr Tanoj Kollamparambil Dr Anitha James	Soha.Elbehery@wales.nhs.uk Sandeep.Ashtekar@wales.nhs.uk Marion.Schmidt@Wales.nhs.uk Chuen.Poon@wales.nhs.uk Tanoj.Kollamparambil@wales.nhs.uk Anitha.james@wales.nhs.uk
Swansea Bay UHB	Singleton	Dr Geraint Morris Dr Maha Mansour	Geraint.Morris@wales.nhs.uk Maha.Mansour@wales.nhs.uk

		Dr Sree Nittur Dr Ankita Jain	Sree.Nittur@wales.nhs.uk Ankita.Jain@wales.nhs.uk
Cwm Taf Morgannwg UHB	Bridgend RGlam Merthyr	Dr Max Nathan Dr Tony Goodwin Dr P Govindaraj Dr Marcia Scheller Dr Rainer Fortner	Max.Nathan@wales.nhs.uk Anthony.Goodwin@wales.nhs.uk Poonamallee.R.Govindaraj@wales.nhs.uk Marcia.Scheller@wales.nhs.uk Rainer.Fortner@wales.nhs.uk
Hywel Dda UHB	WWGH Withybush	Dr Prem Pitchaikani Dr Sian Jenkins Dr Faumy Hassan Dr Debasis Biswas	PremKumar.Pitchaikani@wales.nhs.uk Sian.Jenkins14@wales.nhs.uk FaumyMHassan.MohamedHassan @wales.nhs.uk Debasis.Biswas@wales.nhs.uk

1.6 Annual and Study Leave

- Full leave entitlement should be taken – this needs to be coordinated between the junior team
- Unless there are exceptional circumstances (such as examinations or job interviews) two non-consultant doctors must be present to cover the unit during normal working hours.
- Book leave 6 weeks in advance
    - Inform Dr Amos Wong who will go through the leave diary
    - Use Intrepid system for electronic approval

1.7 On-Call Arrangements

1.7.1 Consultant Staff

- The consultant staff work a 1:6 rota, one week at a time, with a hand-over taking place on Monday mornings in the Planning Meeting.
- An on-call rota is circulated well in advance.
- All junior staff should ensure they know which consultant is on call.
- If you are unable to contact the on-call consultant in an emergency, contact one of the other consultants (or, in a dire emergency, the consultant covering the Bristol unit).

1.7.2 Junior Medical Staff

- The SHO participates in the paediatric specialty on-call rota. Cross-cover arrangements are in place with other specialty teams.
- The rotating general paediatrics registrar attached to the cardiac unit ordinarily participates in the paediatric specialties middle grade rota.
- The cardiology registrar is on-call 1 night in 5 (non-resident) and does not participate in the general paediatrics rota; he or she may be asked to provide cross-cover support during an emergency.
- The resident on-call specialty SHO and specialty registrar provide out-of-hours cross cover for cardiac patients except those receiving critical care on the Paediatric Critical care Unit. Clear hand-over between clinical staff is essential. Hand-over rounds take place daily (Mon-Fri) at 08:30 and 16:00. The team-member covering the cardiac patients should attend the relevant hand-over round to pick up and convey any relevant information about the cardiac patients. Written hand-overs should be provided to the paediatric team when they are cross-covering the cardiac patients.

1.8 Audit and Research

Effective clinical audit can improve patient care. The broad principles behind medical audit are the setting of an accepted standard, comparison of current practice to that standard, making alterations and completing the audit by re-assessing the standard of care.

This Department is actively involved in medical audit, both with the Cardiology service and General Paediatrics. Rotating paediatric junior staff should attend the paediatric audit meetings. Each junior doctor will be expected to undertake an audit project during his or her post.
Participation in research projects is expected from all junior staff and opportunities will exist for presentations at audit meetings or to bodies such as the Welsh Paediatric Society and the British Congenital Cardiac Association. The consultants will provide help and advice on projects.

1.9 Computers and Cardiobase®

1.9.1 Protecting Data

- All medical records held on computer are subject to the Data Protection Act.
- You will be provided with a password for accessing the hospital network.
- The departmental clinical lead bears responsibility with the IT Department for use of the network by the junior staff.
- Any abuse will be dealt with in keeping with health board policies.
- Do not divulge your password to non-unit staff.
- Remember to log-off after use, and use screensavers and other security measures.
- Emails bearing patient identifiable data (PID) may be sent to wales.nhs.uk accounts, but not to university or private/commercial email accounts
- Emails with PID being sent to nhs.uk email addresses in England (e.g. to Bristol) should be encrypted (use the Outlook encryption tool), or using NHSnet.
- **Never take non-encrypted electronic patient identifiable information away from the hospital setting.**
- It is essential that care be taken to avoid the introduction of computer viruses to the network in order to protect the integrity of the patient database.
    - Sources of “infection" are unauthorised software, CDs, DVDs, and memory sticks.
    - Before such software or storage media are used they must be scanned for viruses.
- Hospital disciplinary action may ensue if these rules are not observed.

1.9.2 Cardiobase®

The Unit maintains a comprehensive patient database (Cardiobase) which includes details of each patient, the diagnosis, previous surgery, and events such as outpatient consultations and echocardiograms.

- Cardiobase is available to all staff for the extraction of information.
- Data input is reserved for trained individuals – ask one of the regular users.
- Request a username and login from Amanda Doyle or Beth Corris – discuss with them what level of authority you will have – usually this is data entry, but not data “completion”.
- All new patients entered on to Cardiobase should have the following recorded as a minimum:
    - Care record number
    - Full name
    - NHS number
    - Address including postcode
    - Diagnosis
    - Antenatal diagnosis (Y/N)

Access to Cardiobase out of hours: Patient information is held on Cardiobase. The database is accessible out of hours from any hospital PC (using Microsoft Edge). To gain access:

1. Find the Cardiobase icon on C&V business apps Business Apps on CAVUHB Device (sharepoint.com) OR

2. Log-in to any PC and paste this into the URL section: https://Cardiobase.cymru.nhs.uk and press enter. The programme will load.

For generic read-only access, log in as **doctor** (username) **junior** (password). **You are not able to alter the patient record.**

1.10 Medical Notes and Correspondence Headings

Medical notes should always

- Be written legibly in black ink (blue is also permitted, but black is preferred)
- Be accurate and relate to the correct patient
- Have 3 correct elements of the patient's ID recorded on each page
- Have entries which are **dated, timed** and **legibly signed** by the appropriate doctor or nurse relating to each patient contact – many signatures are illegible, so **print your name and ideally your GMC number under the signature**
- Record verbal advice given to patient or relatives
- Record explanation of risks/benefits of proposed treatment explained to patient or relative
- Contain results of investigations and record action taken on abnormal results
- Be contemporaneous - not written days later (and if they are written "after the fact" this should be stated)
- Be capable of being read out in court by the patient's barrister
- At discharge contain a list of all diagnoses including co-morbidities and procedures (get a senior member of staff to confirm the entries), avoiding the use of ambiguous abbreviations

1.10.1 Correspondence Headings

Medical Discharge Summaries
Date of Admission
Date of Discharge
Cardiologist
Diagnosis
Procedure
History/Examination
Investigations
Management
Status at time of discharge
Weight at discharge
Discharge medication*
Follow-up
Risk of endocarditis Y/N

Copies to _____

Inpatient Reports (see Section 2.2)
Date of consultation
Referring Doctor
Cardiologist
Ward
Reason for referral
Pertinent history/examination
Investigations
Final diagnosis
Advice given
Follow-up

Copies to _____

Top copy of form to patient notes
Bottom copy for dictation/secretaries

The service doctor of the day bears the responsibility of dictating/typing the correspondence, which should be dispatched to the clinician/GP by 5 working days.

Outpatient Letters
Date of clinic / dictation
Date of typing
Cardiac diagnoses
Other diagnoses
Medication (+ any changes)*
Findings, including height and weight
Investigations
Communication with parents
Follow-up

Risk of endocarditis Y/N
Copies to _____

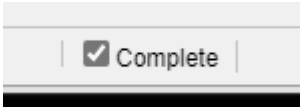
*For CARDIAC medications – these should be stated as the dose to be given (in mg or microgram), and also, set out in brackets, the desired number of mg/mcg per kilogram for this child. Do not state number of millilitres in the letter, as concentrations can change when the prescription is renewed in the community.

Parents are sent a copy of clinic letters. Ensure that the summary at the end of the letter will be understandable to the parents and contains terms a layperson would understand. If the parents are separated/divorced take care not to divulge sensitive, confidential details of the estranged family.

A pro forma Conference Reports is incorporated into the appropriate section on Cardiobase.

Remember to copy relevant correspondence to local consultant or DGH case notes in the case of non-Cardiff residents (see section 1.5 for local doctors with cardiology expertise).

Only letters and reports that are ticked "complete" will population C&V Clinical Portal and Welsh Clinical Portal. Usually the relevant secretary will tick "complete" but can only do so if she is aware of the letter – please telephone or email her to let her know about the correspondence.



1.10.2 Daily Timetable

Monday	08:15	Planning Meeting	Sophie Pearson Room
	09:00	OU and ADP Clinic	Starfish OPD
	12:30	Microbiology and X-Ray Meeting (Paeds)	Paeds Seminar Room or KRUF Seminar Room
	14:00	Nurse-Led Clinic	Starfish OPD
Tuesday	09:00	DGW and VDO Clinic	Starfish OPD
		Fetal Clinic	Antenatal Clinic/FMU
		Adult ASD closures	Cath Lab
		MRI list (non-GA)	Radiology
	14:00	Echo Clinic + Oncology patients	Starfish OPD
Wednesday	08:00	Echo meeting (adult)	C3 Seminar Room
	09:00	Academic Session	Sophie Pearson Room
	10:00	Teaching Ward Round	Pelican + other wards
	12:30	Hospital Grand Round	Lecture Theatre 2
	13:30	CDG and NH clinic	Starfish OPD
	13:30	Adult Congenital Clinic	Cardiology OPD
Thursday	0830	CVS-Genetics MDT	1t Thursdays, SP Room
	09:00	Fetal Clinic	Antenatal Clinic/FMU
	13:00	SHO In-House Teaching	Paeds SemRm
	14:00	Fetal Clinic	Antenatal Clinic/FMU
	14:00	CVS Imaging MDT Meeting	4th Thursdays, SP Room
	15:30	Resp/Cardiac MDT Mtg	4th Thursdays, SP Room
Friday	09:00	Echo Clinic	Starfish OPD
	13:00	Paediatric Case Presentations	Level 3 Seminar Room
		Cardiology Academic Teaching	LHS Pathology Seminar Room
	13:30	Nurse-Led Clinic	Starfish OPD

1.11 Teaching Ward Round

The main round is on Wednesday commencing at 10 am after the teaching session. Each patient is discussed prior to being seen. When presenting the patient's details the following points should be covered:

- Name
- Age
- Diagnosis
- Date of admission
- Reason for admission
- Pertinent social history
- Developmental and immunisation history
- Clinical findings
- Results of investigations
- Latest CXR
- Current the management plan.

All patients must have an up-to-date problem list and growth chart, including head circumference in infants.

Whilst attached to the Department of Paediatric Cardiology, you should take the opportunity to learn the basics of echocardiography (e.g. recognise an effusion). SHOs undertaking MRCPCH examinations need to focus their attention on exam preparation and additional coaching will be provided. A variety of books and journals on congenital heart disease may be borrowed on request, but you must inform the relevant consultant. They must not be removed from the hospital and should be returned promptly.

1.11.1 Teaching Topics

Each rotation will have core teaching topics, including:

- Introduction and induction
- Structural and acquired heart disease
- Cardiac emergencies
- Basic ECG interpretation and arrhythmias
- Fundamentals of echocardiography and care of the echo machine
- Case presentations
- Infective endocarditis
- Surgical treatment of congenital heart disease

Other more specialised topics can be taught on request. NB the teaching sessions are "broadcast" via Microsoft Teams across Wales. Ask to be added to the paediatric cardiology group on Teams.

SECTION 2 – ADMISSIONS AND DISCHARGES

2.1 Categories/Definitions of Admissions and Reviews

Care may be **scheduled** or **unscheduled**. Sources of unscheduled care include:

- The Emergency Department
- The Children's Assessment Unit (referral by GP or paediatrician)
- Direct GP referral
- Self-referral via specialist nurse contact or out-of-hours call to the ward
- Clinic attender needing urgent care
- Casual ward attender where a decision is taken to admit

Category	Explanation	Action
Admission	Patient is admitted to a bed. Nursing resource is used. The patient stays overnight at least one night.	Dictate or type a "Discharge Summary" on Cardiobase. Enter any procedures on the "cardiac admission pro forma".
Day Case Admission	Patient is admitted to a bed for an investigation or procedure (e.g. Captopril challenge, sedated echo). Nursing resource is used. The patient is discharged the same day.	Dictate or type a brief "Discharge Summary" on Cardiobase. Enter any procedures on the "cardiac admission pro forma".
Casual Ward Attender	The patient attends the ward without being admitted to a bed (e.g. blood test).	Dictate or type a "Clinical Letter" on Cardiobase if treatment is changed or if the outcome of the visit needs to be communicated to the GP/DGH.
Outpatient Visit	A patient seen in a booked outpatient clinic (including Nurse-Led Clinic).	Dictate an "Outpatient Letter" or "Nurse-Led Clinic" letter on Cardiobase.
Inpatient Referral	A patient is referred by another team for review whilst they are an inpatient. This may be at UHW or another hospital.	Dictate or type an "Inpatient Report" on Cardiobase. Inpatient reports should also be dictated following a teleconsultation.

2.1.1 Actions for all Admissions

Ensure that the Paediatric Flow Manager (Bleep 6434) is aware of the admission.
Planned admissions need to be entered into the Ward Diary. The entry needs to include:

- Name of patient + case record number
- Diagnosis
- Consultant or specialist nurse requesting admission
- Reason for admission – state if this is "day case" or "admission for overnight stay"
- Name of person making this entry

All admissions receive a pack on arrival in Pelican Ward that includes general information about the Unit, hospital procedures, layout and benefit entitlements. A routine paediatric clerking is required for all admissions. Developmental status is important, as neurological problems noted post-operatively

may have been present pre-operatively! New patients with dysmorphic features or complex disease should have genetic analysis, renal and cranial ultrasound and a genetics referral.

You must document:

- Diagnosis
- Previous cardiac catheters and operations
- Social, developmental, vaccination, coagulation history (excessive bleeding / bruising / history of thrombosis)
- Clinical status and exercise ability
- Height, weight and head circumference (**plot these on a centile chart**)
- 4-limb blood pressure (all new patients)
- Oxygen saturation
- ECG - arrange if one has not been performed in the past three months, or if any planned treatment could lead to ECG changes
- Chest X-ray - consider the need (mandatory for post-op transfers in and patients with an oxygen requirement)
- Echocardiogram findings
- Action plan and problem list
- Results of any investigations undertaken

2.2 Unscheduled Care

2.2.1 Telephone Advice

The Unit operates an "**open access to advice**" policy for all known patients. During working hours most calls are fielded by the cardiac specialist nurses or consultants (via their secretaries). Out-of-hours, parents may contact the ward and the advice of junior staff will usually be sought. If this happens, judge whether the patient should be seen urgently in the Emergency Department, in the CAU by the Paeds Cardiology team (cardiac problem) or by their GP and referred to the Children's Assessment Unit (e.g. for general paediatric problems). Communicate the decision with all relevant parties. If the patient is seen in CAU for a cardiac problem, the consultant on-call should be informed **whatever the time day or night**.

If telephone advice is given, please use the "Telephone Advice" pro forma kept on Pelican Ward, or print out a sheet from the folder on the shared directory (S:\PaedCard\shared\TELEPHONE ADVICE).

2.2.2 Referrals

New patients will be referred to the Unit from a variety of sources (e.g. clinic, GPs, paediatricians etc.). The consultant may prefer to see neonatal referrals at the cot side in the referring hospital, so check before arranging transfer to UHW. Many neonates, once seen by the UHW cardiologist, are referred directly from the referring hospital to the surgical centre.

For calls from outside of Cardiff:

- Ascertain the basic history and clinical status of the patient from the referring doctor (e.g. age of child, oxygen saturation, presence of cardiac failure, ventilation status, and overall condition).
- The name and telephone number of the referring doctor should be noted and then the cardiologist contacted.
- Remember to contact the referring hospital when the cardiac condition is established and update them on the diagnosis and treatment plan.

Inpatient referrals from UHW/Children's Hospital are frequently received from neonatologists, paediatric surgeons and general paediatricians.

- Before reviewing the patient, ensure the consultation has been requested by/discussed with the referring consultant.
- **It is essential to clarify the clinical question** being posed by the referring team.
- If the question is not clear, discuss this with the cardiac consultant on-call.
- **Referrals made after 3 pm: the referring team needs to discuss the referral with the on-call cardiac consultant.**
- These patients should have an outpatient-style consultation, a full CVS examination and an ECG, echocardiogram, and chest x-ray (where appropriate, e.g. if the saturations are abnormal or if there are features of heart failure).
- The findings should be discussed with the on-call consultant, preferably well before the end of the normal working day – remember all echos are reviewed by a consultant.
- Details of the consultation should be documented in the case record notes, including information about the echocardiogram, and an **inpatient report** dictated or typed.
- Telephone or email the on-call consultant's secretary to inform her that the letter needs to be tidied up, **ticked as completed** and sent to all relevant parties.
- Ensure copies of this report are sent to the parents, the GP and the patient's DGH (since the parents are receiving a copy, ensure the summary at the end is in plain English).
- Inpatient referrals should be reviewed as necessary and follow-up arrangements should be clearly stated.

NB – There is almost no such thing as a referral for "echo only". Exceptions are follow-up neonatal echos (check known PDA), or chemo patients having LV function assessment between courses (if there have been no previous problems). All other echo requests are regarded as a "referral for cardiology opinion".

Inpatient referral checklist:

- Was the referral generated by a consultant?
- Is the clinical question clear?
- Take a history and perform a clinical examination
- Arrange essential investigations (consider need for CXR)
- Are the findings adequately documented in the medical record?
- Has the case been discussed with the consultant?
- Has the clinical problem been addressed?
- Has the outcome been clearly communicated to the referring team/GP/DGH paediatrician?
- Has an inpatient report been typed or dictated, checked, **ticked as completed** and dispatched, including to the DGH if the patient is not a Cardiff resident? **Have you spoken to or emailed the relevant cardiac secretary?**
- Are any outpatient arrangements in place?

See Administration section for Inpatient Report headings (1.10).

2.2.3 Neonatal Referrals

Ensure the birth weight and gestational age are documented in the medical records and on Cardiobase.

Special considerations apply to the management of neonates with suspected congenital heart disease as many will have duct dependent lesions and some will have single ventricle physiology. The history and examination are important in distinguishing cardiac from respiratory causes. Investigations should include the pre- and post-ductal O_2 saturation, 4-limb BP, chest X-ray, ECG, FBC, U&E and capillary blood gas + lactate (if unwell).

Many will have duct dependent lesions and there should be a low threshold for starting prostaglandin. Even despite this mixing may be inadequate and urgent balloon atrial septostomy may be necessary.

Those with single ventricle physiology (e.g. HLHS) will require a different ventilation strategy and babies with dysmorphic features (e.g. DiGeorge Syndrome) irradiated, CMV negative blood products.

2.2.4 Casual Ward Attenders

Patients attend the ward for a number of reasons, including post-transplant supervision, INR estimation, post-operative review, parental concern, and so on. Some of these patients can be reviewed in the Nurse-Led Clinic to avoid burdening the ward nurses. The patient's diagnosis, active problems, weight, and current progress should be noted. If there are any concerns then the registrar or consultant should be informed. Any significant changes in therapy or management should be communicated with the patient's GP by typing or dictating a Clinical Letter on Cardiobase (inform secretary)

2.2.5 Post-Surgical Transfers In

Following cardiac surgery some cases are transferred back to Cardiff prior to final discharge home. This is usually because their postoperative course is slow or complicated. They may have:

- Feeding problems
- Persisting haemodynamic problems
- Pericardial effusion
- Persisting respiratory problems
- Persisting neurological problems
- Ongoing infection

See the relevant section in Clinical Problems or Surgical Complications. All patients transferred to Cardiff for post-operative management need an up-to-date ECG, CXR and echocardiogram.

2.3 Scheduled Care

Patients are admitted electively to Pelican for a number of reasons:

- Day case investigations or treatment
- Admission for CT or MRI under GA (see “Investigations” section)
- Observation
- Monitoring
- Cardiac input into an admission for non-cardiac care, e.g. dental treatment, plastic surgery or ENT procedures. The main reason is to provide cardiac supervision during the admission and allow anaesthesia in main theatres with full cardiac back up. The NICE guideline for preventing infective endocarditis should apply – unless specified by the consultant (e.g. very high risk cases) no antibiotic cover is given.

2.3.1 Dental Admissions

Patients with important underlying cardiac disease may be admitted to Owl/Gwdihw Ward or Pelican for medical oversight of their admission. The dental team will arrange for a dental review + consent. The cardiology team needs to document a brief history and examination and ensure a recent ECG and echo have been performed (e.g. within the last 3-6 months) – if they have not, they will need to be undertaken and the findings documented.

2.4 Transfers and Discharges

General principles

- Before transferring or discharging a patient, imagine yourself as the doctor receiving the transfer, or the paediatrician in the DGH, the GP, or the consultant seeing the patient in clinic in a few weeks’ time. Also picture yourself as the parent taking their child home after a stressful time in a specialist centre. Have they received clear communication about the status of their child and the management plan? Do they know whom to contact if they are worried?
- Clear verbal and written communication is paramount.
- Provide a Discharge Advice Letter via Welsh Clinical Portal – reference the fact a full summary will be sent subsequently.
- Sufficient drugs should be prescribed for up to one month (if needed that long).

- Parents should be told to contact their GP and local pharmacy soon after discharge for a repeat prescription as many of the drugs commonly prescribed from hospital (e.g. furosemide suspension) need to be specially ordered and the chemist will require a few days to obtain them.

It is the responsibility of the SHO or registrar on-call the day of the patient's discharge to ensure a summary is dictated that day. **See section 1.10 for a guide to discharge summary headings.**

Summaries on patients with protracted or complicated admissions should be dictated or checked by one of the registrars. As a minimum standard, all discharge summaries should be dictated, typed and posted within five working days of discharge from the hospital.

2.4.1 Transfers to Other Hospitals

Before transferring a child to the surgical centre consider what level of transfer should be undertaken.

- Always use the Inter-Hospital Transfer Document (S:\PaedCard\shared\TRANSFERS).
- Always phone ahead to ensure the receiving hospital still has a bed.
- Well patients may be transferred safely in the family car or in a hospital taxi with a nurse escort.
- Patients on minimal support (e.g. intravenous drugs given by bolus, patient receiving low-flow oxygen) may be appropriately transferred by the nursing staff.
- Patients receiving a higher level of support (e.g. prostaglandin infusion) must be transferred with an experienced member of the medical staff with a paramedic crew. Intubated patients will normally be transferred under the supervision of NICU or PICU consultant staff.
- A written summary (typed wherever possible) should be provided and this should detail all of the relevant medical details, including an up-to-date medication list.
- In some circumstances a photocopy of the patient's medical notes should also be sent. If time constraints do not permit a typed summary, a clear hand-written note is required, **but a summary should be dictated for typing later**. This permits accurate audit of transfers out of the Unit.

Emergency Transport Services

Neonatal emergency transfers are overseen by the CHANTS Team. CHANTS operates a 24/7 service, 365 days a year. The service rotates between Cardiff, Newport and Swansea, working 1 week in 3 in rotation.

Contact numbers are:

Swansea – 01792 285278/5403 **Cardiff –** 02920 744719 **Newport –** 01633 234844

Paediatric emergency transfers are overseen by the WATCH team. WATCH is staffed 24 hours a day, seven days a week, by a specialist paediatric intensive care consultant, registrar, advanced transport nurse practitioners and transport nurses. The team offers advice, accept referrals and transfer critically ill children back to the appropriate paediatric intensive care unit. It is also commissioned to provide HDU-HDU transport.

The contact number is: 03000300789.

Transfer Checklist

- o Is there a clear indication for transfer?
- o Ensure patient is fit for transfer – start to fill in the Inter-Hospital Transfer Document
- o Communicate with medical staff in the receiving unit
- o Ensure appropriate ambulance has been arranged
- o Ascertain which members of the medical/nursing team should accompany the patient – see interhospital transfer document for guidance
- o Written transfer letter (preferably typed)
- o Document current medications and recent lab results
- o Should any of the notes be copied?
- o Ensure the proper equipment is available (transfer pack)

- o Ensure safe level of monitoring equipment
- o Telephone receiving hospital just prior to departure
- o Ensure a discharge summary is dictated/typed (inform secretary)

2.4.2 Pre-Operative Checklist

If a patient is being worked up / transferred for urgent cardiac surgery, please ensure the following is in place:

- Liaison with surgical coordinator in Bristol to confirm date of surgery (0117 342 8862)
- Ensure pre-op surgical swabs are sent:
  - o Throat and nasal swabs for MRSA and MSSA – see Section 6.17 if these come back positive
  - o The request form must specify "Bristol pre-op screening protocol"
  - o It is advisable to telephone the lab to ensure the samples are run as requested
- Determine how the patient will get to Bristol – see inter-hospital transfer documentation (above)
- Ensure the family knows what to expect when they get to Bristol – involve the cardiac nurse specialists.

2.4.3 Discharges Following Cardiac Surgery

- The standard hand-written GP note should be provided and the post-surgical handout for the parents should also be given.
- Where the post-operative course has been prolonged or complicated, the DGH consultant and GP should be informed of the discharge by telephone.
- A typewritten summary should be dispatched within five working days.

Wound Care and Physical Activity:

- Normal bathing is permitted but the wound should be protected until the scabs have come off.
- Parents should know that if the child is unwell prior to the OPD appointment, the ward should be informed and a review will be arranged.
- Most children should not attend school until after their first review in Nurse-Led Clinic or OPD. As a general rule the child should avoid PE (or equivalent activity) for 6/52 and contact sports (or equivalent) for 3/12. Other disease-related activity restrictions may apply.
- Follow-up Arrangements: 2 week appointment Nurse-Led Clinic, 4-6 week appointment Cardiology OPD (at local hospital where possible) – **ensure these arrangements are in place**.

2.4.4 Discharge Checklist

- o Is the patient fit for discharge?
- o Are the medical notes complete with full documentation of the patient's status?
- o Has there been adequate communication (with parents, nursing staff, GP, referring paediatrician?
- o Is the Discharge Advice Letter and TTH complete?
- o Are suitable follow-up arrangements in place?
- o Has the discharge summary been dictated/typed with copies to relevant individuals, including the referring DGH consultant?
- o Has the relevant secretary been informed so that she can tidy up and tick the letter as completed?

2.5 Death of a Patient

Inform the on-call consultant immediately and the child's consultant, even if not on-call, at the earliest opportunity. Consider the need for the coroner's office to be informed of the death. It is Unit policy to

ask the family to consider a hospital PM (either full or limited) if the death is not a coroner's case. Use the new hospital PM consent form.

Contact:

- GP and Health Visitor
- DGH Consultant
- Birth Clerk/Community Paediatrics (to prevent routine appointments being sent to Cardiff patients)
- Cardiac Nurse Specialist – ask her to liaise with the Cardiac Clinical Psychology team – see what bereavement support can be put into place
- Consultant Obstetrician (in cases of neonatal death)
- Consultant Cardiac Surgeon (in the case of post-operative death)
- If the death was unexpected, inform the Clinical Director, enact the hospital PRUDIC protocol and ensure the death is reported to the WPSU Child Death Review process.

https://phw.nhs.wales/services-and-teams/national-safeguarding-team-nhs-wales2/safeguarding-latest-guidance/specific-groups-accordion/prudic-procedural-response-to-unexpected-deaths-in-childhood/

As the notes are usually "lost" in the pathology department after death a summary should be dictated at the time the death certificate is signed. Copies of this should be sent to everyone who has been involved in the patient's care, including previous hospitals. If not all the information is available for the summary at this time explain an additional report will follow.

A leaflet is available on the ward that gives advice to relatives of the deceased about obtaining the death certificate and contacting funeral directors. The patient's consultant will normally write to the family of the deceased child a short time after the event to see if they want to discuss the death of their child further.

SECTION 3 – CLINICS

3.1 UHW Clinics including Pacing Clinic

3.1.1 General Clinics

The outpatient setting will provide good experience in listening to murmurs and dealing with some of the long-term management issues in paediatric cardiac patients. Make use of these clinics by attending regularly.

3.1.2 Specialist Clinics

Experience is also offered in the specialist clinics, including Marfan, Pacing, Adult Congenital and Fetal clinics.

3.1.3 Pacing Clinic

See also:
Emergency Procedures Section *4.4 Temporary Pacing in Children*, and
Clinical Problems Section *6.20 Pacing in Children and Adolescents* for further information.

The Pacing Clinic is held monthly. It is overseen by the Speciality Doctor and the Cardiff Paediatric Pacing Lead Consultant.

Referral into the Service

Most new patients will be referred following pacemaker implant in Bristol. The Bristol Pacing Clinic will undertake the first pacing check at 6 weeks. Immediately after that visit the Bristol team will send to the UHW Device Team and the Cardiff Paediatric Pacing Lead Consultant the following:

- Copy of the implant form
- Latest interrogation data

In patients for whom remote monitoring is required, a referral will be made, as above, from the Bristol to the Cardiff team. Additional information to be transferred will include the serial number of the device and/or the transmitter. This information may be transferred electronically or in paper format depending on the manufacturer.
The vast majority of patients will receive long-term pacing follow-up in Wales.

Patients "Retained" in the Bristol Pacing Clinic

Where there is a clear clinical reason for ongoing review with the Bristol service, copies of implant data and all correspondence will be sent to the UHW Device Team and the Cardiff Paediatric Pacing Lead Consultant. This will ensure up-to-date information is available in Cardiff in case of local emergencies.

General Cardiology Follow-Up

All patients with a pacemaker or device, whatever the indication, will have a named paediatric cardiologist who will oversee their general care. It is expected that all paced patients will have echocardiography to assess ventricular function on at least an annual basis.

Pacing Clinic Visit

All patients:

- Ht }
- Wt } taken by
- Saturations } POPD nursing staff
- Blood pressure }
- 12-lead ECG by ECG technician
- Review need for 24 hour tape
- Review the need for an exercise test

- Review need for CXR (e.g. if rapid increase in height/weight – to be arranged by specialty doctor or trainee overseeing the clinic)
- Full pacing check by pacing cardiac physiologist (PCP)
- Data input directly by PCP and staff grade on to new pacing module on Cardiobase with report generated immediately for (a) medical notes, (b) pacing file, (c) GP, (d) named consultant (e) DGH notes (for non-Cardiff residents). The pacing clinic letter will make it clear who was present and participating in the clinic.

Definition of "high-risk" devices with mandatory consultant review:
- Cardiac resynchronisation therapy pacing
- Automated implantable cardiac defibrillators
- Any pacemaker with evidence of battery depletion or a lead problem
- All infants/children <2 years of age

All relevant results will be conveyed to the Cardiff Paediatric Pacing Lead Consultant and the patient's named consultant.

The Paediatric Pacing Clinic runs on a monthly basis. All parties need to be informed if there is any change in the clinic date. If the Cardiff Paediatric Pacing Lead Consultant is away, the on-call (or covering) paediatric cardiology consultant will take responsibility for overseeing "high risk" cases.

Implantable Loop Recorders (Reveal devices, LINQ)
Bristol will inform the UHW Device Team and the local consultant paediatric cardiologist when such a device is implanted. All patients presenting with symptoms will be assessed by the Cardiff pacing team and the device will be analysed. The results will be conveyed to the local consultant paediatric cardiologist.
Contributors: Reviewed 2023 by Amos Wong, Chris Gillett, Andy Penney and Georgia Spentzou

3.1.4 Echo Clinic
The paediatric cardiac speciality doctor undertakes an "Echo Clinic" twice weekly. Referrals to the clinic are made via the consultant medical staff. The on-call paediatric cardiologist provides consultant support for this clinic. The following types of referral are seen in this clinic:
- Oncology patients having routine inter-treatment scans where the question is ?LV function
- Nephrology patients where the question is ?LV function, ?LV hypertrophy or those pre- or post- renal transplant needing "routine" echocardiography
- Patients seen by or discussed with a consultant general paediatrician with a murmur where the clinical assessment is that it is innocent and confirmation by echo is desired (patients where pathology is suspected should be referred to a formal paediatric cardiology clinic)
- There is weekly provision for a Rapid Access Clinic slot – this needs to be discussed and approved by the speciality doctor running the clinic.

3.2 Nurse-Led Clinic
- Nurse-Led Clinic runs on Mondays 14.00 – 16.30 and Fridays 13.30 – 16.30 in the Starfish Outpatient Department.
- The clinic is supported by the cardiac dietician and echo physiologist; junior doctors may be asked to review patients attending the clinic.
- Aspects of care provided include
    - Post-operative review
    - Nutritional review
    - Review of patients between cardiac outpatient clinic visits
- Patients are booked by an appointment system allowing 15-30 minutes per patient.
- A diary for these appointments is kept in the Liaison Nurses' Office.

- The person booking the appointment is responsible for informing the patient/parents of the date, time and location of the clinic.

3.3 Outreach Clinics

UHW consultants undertake >200 peripheral clinic sessions each year in each local health board (LHB). If you make the effort to attend some of these clinics you will be rewarded with a wealth of training opportunities. Post-discharge patients should be slotted into one of these local clinics, where appropriate – check with the consultant.

Consultant responsibilities for LHBs

LHB	Hospital	Consultant providing input	Sessions provided p.a.
Aneurin Bevan	Nevill Hall Hospital (Abergavenny)	CDG, DGW	CDG 36 DGW 48 OU 12
	St Woolos Hospital (Newport)	CDG, OU, DGW	
Cwm Taf Morgannwg	Prince Charles Hospital (Merthyr)	VDO	VDO 30
	Princess of Wales Hospital (Bridgend)	OU	OU 40
	Royal Glamorgan Hospital (Talbot Green)	ADP, NH	ADP 12 NH 20
Hywel Dda	Glangwili General (Carmarthen)	NH, VDO, DGW	NH 24 VDO 12 DGW 4
	Withybush General (Haverfordwest)	NH, VDO, DGW	VDO 12. DGW 4
Swansea Bay	Morriston Hospital	NH, VDO, ADP	NH 28 VDO 24 ADP 24

Transition clinics are held in each LHB 2-4 times a year depending on need.

ACHD clinics are also held in each LHB.

SECTION 4 - EMERGENCY PROCEDURES

4.1 Balloon Atrial Septostomy

BAS may be required to improve mixing between the pulmonary and systemic circulations in conditions such as:

- Transposition of the great arteries
- Tricuspid atresia
- Pulmonary atresia with intact septum.

In most cases BAS may be performed semi-electively in the surgical centre, but in many cases it is a life-saving procedure in a collapsed neonate. In most circumstances BAS is performed with sedation and local anaesthesia ± muscle relaxant in a ventilated neonate. In experienced hands BAS is usually a very safe procedure, but there is a risk of serious complications including:

- Bleeding
- Pericardial tamponade
- IVC avulsion
- Mitral valve damage
- Stroke
- Arrhythmia.

The risk is higher in certain situations (e.g. in the context of juxtaposed atrial appendages, atrial septal aneurysm, or interruption of the IVC). The risks of the procedure must be weighed against the risk of delaying life-saving intervention. Except in life-threatening situations, high risk BAS should be delayed until transfer to a surgical centre.

In dire emergencies, the on-call Bristol interventional consultant may opt to undertake emergency septostomy in a Welsh hospital. They will bring a septostomy kit with them.

Prerequisites for BAS

- Skilled operator
- Support staff trained in safe airway management (this may be an intensivist, neonatal consultant or an experienced NICU registrar)
- Safe environment with full monitoring and resuscitation equipment (e.g. PICU or NICU)
- Equipment necessary for the procedure (brought by Bristol consultant)
- 2 units of cross-matched blood
- Informed consent from the parent/guardian

Contributor: Dirk Wilson

4.2 DC Cardioversion (DCCV)

Patients with a shockable rhythm in cardiac arrest should receive DCCV as part of the resuscitation algorithm (this is undertaken wherever the arrest is being managed and does not require separate sedation or anaesthesia).

DCCV outside the context of cardiac arrest may be needed in cases of shockable atrial or ventricular arrhythmia with cardiovascular compromise, e.g. rapid SVT not responsive to medication, VT with a pulse but with features of shock, or where semi-urgent DC cardioversion is needed as “first-line” therapy, e.g. atrial fibrillation or flutter.

Prior to carrying out DCCV, ensure the following:

- The patient should be nil by mouth (the “fasting 2,4,6” rule applies except in dire emergency: NBM 2 hours for clear fluid, 4 hours for breast milk and 6 hours for formula milk or solids)
- Inform the on call paediatric anaesthetist
- Inform Paediatric Theatres (47457 / 47337) or Main Theatres (42993) if that is the preferred location for the procedure
- Inform PICU (in case admission is required afterwards)

- Obtain formal consent. The potential risks include:
  - Intractable cardiac arrest
  - Neurological deficit
  - Surface irritation or burns to skin
  - Risks of anaesthesia

Procedure:

- Once the patient is anaesthetised and stable, set up the defibrillator as per APLS/PLS guidance.
- Ensure you are recording the process on paper (i.e. ECG running).
- Mark events on the machine if this facility is available.
- Apply appropriate energy

Synchronised cardioversion
With appropriate sedation + analgesia (e.g. IM/intranasal Ketamine if delay in IV access + airway management) – IV access attempts must not delay cardioversion **1st shock:** 1 J kg^{-1} **2nd shock:** 2 J kg^{-1}, consider up to 4 J kg^{-1}

If an energy level of 2 J/kg is unsuccessful, consider changing pad position to anterior-posterior (apex and L subscapular), or reverse polarity (reverse position of apex and subclavicular paddles). Consider using up to 4 J/kg.

After DCCV, admission to PICU should be considered if the patient remains haemodynamically unstable – have a low threshold if ventricular function is poor. Consider what prophylactic anti-arrhythmic medication is required.

Contributors: Orhan Uzun and Dirk Wilson

4.3 Pericardiocentesis for cardiac tamponade

Aetiology

Cardiac tamponade may accompany a variety of illnesses, such as viral infections (e.g. coxsackie, mumps, adenovirus, and HIV), bacterial infection (e.g. TB), immune mediated diseases (e.g. rheumatic fever, Kawasaki disease), connective tissue diseases (e.g. JRA, SLE) and uraemia.

Pericardial effusion is also a common post-operative complication. It usually resolves spontaneously or following treatment with diuretics ± anti-inflammatory drugs such as aspirin or ibuprofen. Often the effusion is a feature in the early post-operative period and patients are discharged from the surgical centre only when it is clear the collection is not increasing in size. In other cases the effusion may only become apparent many days or even weeks post-operatively and forms part of the post-cardiotomy (Dressler's) syndrome.

Clinical features

- Prodromal illness
- Chest pain
- Disappearance of a previous pericardial rub
- Tachycardia, tachypnoea, gallop

- Hepatomegaly and raised JVP (evidence of elevated venous pressures)
- GI upset
- Pulsus paradoxus
- Muffled heart sounds
- Cardiac arrhythmia

Investigations
ECG: low voltages, ST-T changes
CXR: cardiomegaly
Echocardiogram:

- 2D:
    - Presence of pericardial effusion
    - Diastolic collapse of right ventricle
    - IVC dilatation and loss of respiratory variations
    - Respiratory increase of inter-ventricular dependence
- Doppler:
    - Respiratory variations > 25% in mitral, aortic and/or tricuspid flow

(see http://www.stanford.edu/group/ccm_echocardio/cgi-bin/mediawiki/index.php/Tamponade) **)**

Management
Post-operative pericardial effusions are monitored with careful clinical and echocardiographic examination.
Patients with features of tamponade or impending tamponade require urgent pericardiocentesis.

Faced with a patient with tamponade/impending tamponade, the clinician must judge whether it is safer to:
(a) perform a pericardial tap locally, or
(b) transfer the patient to the surgical centre for pericardiocentesis.

The potential risks of inadvertent myocardial or coronary damage must be weighed against that of cardiac arrest during transfer. As the worst-case scenario is the need to perform urgent pericardiocentesis during transfer, the patient should be retrieved by the WATCh Team. Except in dire emergency, the patient should be nil by mouth ("fasting 2, 4, 6" rule applies).

Technique
The removal of even a small amount of fluid from the pericardial space can be life saving and attempted relief of tamponade is preferable to the consequences of non-intervention. In an emergency any large intravenous cannula can be used. Clean the xiphoid and subxiphoid areas.

Echo guidance is recommended
Use local anaesthetic if necessary
Attach the syringe to the needle
Puncture the skin 1-2 cm inferior to the left side of the xiphoid junction at a 45^{o} angle
towards the tip of the left scapula aspirating all the time

When fluid is withdrawn advance the cannula over the needle and withdraw the needle
Remove as much fluid as possible
If the fluid is bloody then it can be difficult to decide if the needle is in the pericardial space or heart as the blood will pulsate out of the needle even if it is in the correct place. If the fluid is squirted onto a white swab it is often obvious if the blood is fresh or old. When echo is available a saline can be injected through the needle and will readily demonstrate if it is in the pericardial space or heart

If the cannula is in the correct place the clinical condition of the patient will rapidly improve as fluid is withdrawn.

Prerequisites for Pericardiocentesis

- NBM (2,4,6 rule unless dire emergency)
- Skilled operator
- Anaesthetist or intensivist and appropriate support staff
- Safe environment with full monitoring and resuscitation equipment (e.g. operating theatre, PICU)
- Equipment necessary for the procedure
- Ability to admit/transfer to PICU should complications or the patient's clinical condition dictate
- 2 units of cross-matched blood
- Informed consent from the parent/guardian

Possible scenarios

1. Well child with increasing significant pericardial collection with no signs of response to treatment. Appropriate management – admit to ward for close observation and IV diuretics (if indicated); consider need to semi-urgent transfer to surgical centre for pericardiocentesis.

2. Child with significant pericardial collection and impending collapse due to tamponade. Appropriate management – consider balance of risks of transfer vs local treatment. If the decision is to transfer If a decision is taken to relieve the tamponade at UHW, arrange for transfer to Main Theatres or PICU for urgent pericardiocentesis (paediatric anaesthetist / intensivist support).

3. Collapsed child with evidence of cardiac tamponade. Appropriate management – perform pericardiocentesis at bedside as part of resuscitation (monitored bed). Transfer to PICU for post-arrest management. **Contributor: Dirk Wilson**

4.4 Temporary Pacing in Children

Patients with symptomatic bradycardia may require temporary pacing. The following principles should be applied:

1. Assessment
    a. Undertake ABC assessment – is there evidence of circulatory insufficiency?
    b. Undertake a 12-lead ECG
2. Communication
    a. PCCU consultant
    b. On call paediatric anaesthetist
    c. WATCH team (alert them that a transfer is imminent)
    d. Surgical centre (usually Bristol)
3. Place of safety
    a. ED resuscitation bay
    b. PCCU
    c. Paediatric Theatres or Main Theatres (during pacing wire insertion)

    Usually the patient will move from the ED to PCCU and have the temporary pacing wire inserted on the CEPOD list in Paediatric or Main Theatres with use of an image intensifier – this needs to be agreed between staff at the time
4. Medical therapy
    a. Stop any drugs that may be exacerbating the bradycardia (e.g. β-blockers)

    b. Consider use of isoprenaline to increase heart rate – this may necessitate the placement of a central line – use adrenaline if isoprenaline not available
5. If the clinical judgement is that urgent temporary pacing is required before transfer to the surgical centre, the personnel involved in temporary pacing wire insertion include:
    a. Paediatric cardiologist – advice and support of the on-call adult cardiologist may be sought
    b. Paediatric anaesthetist and intensivist + operating department assistance (ODA)
    c. CCU staff (42110) – ask them (i) to contact the on-call physiologist, (ii) to contact the on call Catheter Lab nurse to provide the 5F Radial Terumo Glidesheath with a small wire along with the 4F balloon-tipped pacing wire, and (ii) ask CCU staff to lay out the temporary pacing trolley with the standard equipment + the special equipment being brought from Cath Lab
    d. Arrange for the temporary pacing trolley to be collected from CCU (Porters or a runner from Theatre)
    e. Theatre nursing staff
    f. Radiographer (for operating the image intensifier – call 43027 during working hours, or 48072 out-of-hours)
6. Insertion of a temporary pacing wire
    a. Sterile technique
    b. Femoral or jugular venous access (ideally ultrasound-guided)
    c. Insertion of a 5F venous sheath (see above) – check wire sizes and work out a suitable “upsize” plan with the anaesthetist
    d. Using image intensifier, pacing wire is advanced to RA, across tricuspid valve and into right ventricle
    e. Physiologist confirms capture and pacing
    f. Set-up pacing box to VVI mode at the desired rate (usually >60-80 bpm)
7. Monitoring
    a. Ongoing heart rate monitoring
    b. Regular observations
8. Return to place of safety: PCCU initially, then transfer to surgical centre – this will involve the WATCh team and the temporary pacing box will need to accompany the patient – ensure the power supply is adequate for the journey.

Contributors: Dirk Wilson, Fiona Macfarlane, Andrew Penney

SECTION 5 – INVESTIGATIONS

5.1 Blood Tests

5.1.1 Drug Levels and Monitoring

See Pathology - Blood-Sciences-test-requirements.pdf - All Documents (sharepoint.com)

Drug	Investigation	Specimen	Comments
Amiodarone	TFT, LFT, CXR and consider Ophthalmology (changes are reversible)	Lithium heparin bottle	Check U&E, TFT and LFT at baseline, at 1 month and then every 6 months. Perform CXR every 12 months. Drug levels (+ metabolites) can be checked through Toxicology Lab, but rarely indicated.
Bosentan monitoring	FBC plus LFT (*must specify ALT and AST*); if on diuretics or ACE inhibitor, do U&E/creat twice yearly (in June and December)	Lithium heparin bottle plus EDTA for FBC	Transaminases may become elevated on bosentan. Bloods done monthly for 4 months, then quarterly if they were satisfactory. Email results to the PAH team at GOSH
Ciclosporin	Drug level	EDTA bottle	Desired range varies – check with transplant team. Email results to the Transplant team at GOSH
Clexane	Anti-factor Xa level	Blue top / paediatric coagulation bottle filled to the line – agitate gently	Liaise with lab particularly if not being taken on “standard” days – see all-Wales protocol.
Digoxin	Drug level	Plain/clotted sample	The level is taken 6-8 hours post dose Desired level = 0.5-2.0 μg/L Check U&E at same time. Consider need for ECG if level abnormal.
Flecainide	Drug level and ECG	EDTA sample (purple) – send at least 1 mL	Trough level at least 6 hours after last dose. QRS duration on ECG should lengthen by <25% of baseline. Desired level = 0.15-0.9 mg/mL (Cardiff Lab – Bristol lab uses range 200-800 μg/L
Tacrolimus	LFT, U&E, FBC and drug levels (plus any other tests specified by transplant centre)	Specimen bottle for Tacrolimus - EDTA	Desired range varies – check with transplant team (bleep 0600 via GOS Switchboard for dosing advice). Fax results to GOS 0207 813 8440

Contributors: Amos Wong, Gareth Davies

5.1.2 Genetic Testing

Blood test required: 2 mL EDTA blood (adult bottles preferred). 0.5 mL of lithium heparin can also be useful if follow up FISH testing might be helpful, but is not essential.

SNP-Array

The standard first line genetic test for CHD (congenital heart disease) is an array. Wales now uses a single nucleotide polymorphism array (SNP-array). This has replaced karyotype and FISH (fluorescence in situ hybridisation) testing.
(https://medicalgenomicswales.co.uk/images/awmgsdownload/PD-GEN-INFSNPArray2.pdf).

The array leaflet gives details of the criteria in the context of:

- Developmental delay / intellectual disability (ID)
- Dysmorphism
- Seizure disorder
- Multiple congenital abnormalities (MCAs)

A cardiac anomaly in the context of one of the above would normally meet the criteria.

An array will detect microdeletions and microduplications (copy number variants, CNVs) across the whole genome in a single test. It will not detect

- Balanced translocations
- Variants in a gene if the normal number of copies are present.

The results may show:

- No CNVs
- Clinically significant CNVs
- Variants of uncertain clinical significance (VUSs)
- Incidental findings (IFs).

It is therefore important to involve the AWMGS (All-Wales Medical Genomics Service) team when interpreting the results in a clinical context. It is also important to note that a normal array does not mean that there is no genetic cause. It may be that there is a CNV (copy number variant) below the resolution of the array, or that there is a causative variant in a gene sequence.

Counselling and consent for genetic testing:
Parents need to sign consent on the request form.
Key counselling points when seeking consent for an array:

- If a VUS is detected, parental testing may be advised to see if it is new in the child – input from the clinical genetics team may be advised.
- IFs may rarely occur and indicate a predisposition to another disease.
- A negative diagnostic result does not exclude a single gene cause of the condition - it may be e.g. that the causative variant is in parts of the genome that we do not yet test.

Genetic testing in Wales has aligned to the NHS England test directory – see
(https://www.england.nhs.uk/publication/national-genomic-test-directories/).

Targeted micro-array testing

Advised testing for CHD patients for whom there is a clear cardiac or dysmorphic phenotype:

Test advised	**Indication**	**Comment**
R137	Conotruncal abnormalities, CHD with cleft palate ± disorder of calcium homeostasis	State on request form, “meets R137 test criteria and specify lesion, e.g. ToF DiGeorge Interrupted aortic arch

		This is a microarray test
R26	Coarctation with features of Turner syndrome	Common aneuploidy test
R27	Congenital malformation and dysmorphism syndromes	Likely monogenic (e.g. **)
R89	Congenital malformation and dysmorphism syndromes	Ultra-rare and atypical monogenic disorders (e.g. **)

If the patient does not meet R137 or the Wales array criteria, you should seek approval at one of the cardiac genetics MDT (multi-disciplinary team) meetings. These happen normally at 0830 every 1st Thursday of the month in south-east Wales in the Sophie Pearson Room in Cardiff or on Teams, every six weeks or so in south-west Wales, and also in north Wales. They are also an educational opportunity - do ask about them. If you need an array more urgently that does not meet criteria, contact the on call clinical genetics team (029218 42577 or se.genetics@wales.nhs.uk) - normally this would be a senior or senior trainee decision.

Gene panel testing

There are other diagnostic tests available for inherited (or inheritable) cardiac conditions (ICCs). The following gene panels are also in the cardiology section of the Test Directory:

Panel	**Testing for**
R125	Thoracic aortic aneurysm or dissection
R127	Long QT syndrome
R128	Brugada syndrome and cardiac sodium channel disease
R129	Catecholaminergic polymorphic VT
R130	Short QT syndrome
R131	Hypertrophic cardiomyopathy
R132	Dilated and arrhythmogenic cardiomyopathy
R391	Barth syndrome
R133	Arrhythmogenic right ventricular cardiomyopathy
R135	Paediatric or syndromic cardiomyopathy
R136	Primary lymphoedema
R138	Sudden unexplained death or survivors of a cardiac event
R328	Progressive cardiac conduction disease
R384	Generalised arterial calcification in infancy
R140	Elastin-related phenotypes

There are other conditions that may involve a cardiac phenotype in other sections of the TD. Diagnostic testing for almost all of these conditions involves a gene panel. Almost all will go outside Wales. All of them have specific criteria listed in the TD. If the patient meets a criterion (and this would normally be a senior decision), you should specify which on the form. If the patient does not clearly meet the criteria, they should have approval in the cardiac genetics MDT meeting. You should then write "approved at MDT meeting dd/mm/yyyy" on the form. If in doubt, bring the case to the MDT meeting.

There are other conditions that may have testing in Wales, e.g. NF1 (neurofibromatosis type 1). These are on https://medicalgenomicswales.co.uk/index.php/download-services including criteria.

Some other conditions no longer have single gene testing available:

- Marfan syndrome - FBN1 testing is only available as part of the panels such as the R125 thoracic aortic aneurysm or dissection panel. Patients need it meet the criteria for the test or have approval at the cardiac genetics MDT meeting

- Alagille syndrome – features on the R171 Cholestasis panel – discuss with clinical genetics
- Noonan syndrome without cardiomyopathy – discuss with clinical genetics.
- Metabolic conditions [e.g. GSD II (glycogen storage disease type II, acid maltase / Pompe disease)] have relevant panels.

Diagnostic testing means the patient already has clear clinical features of the condition and you are looking for the cause. This type of testing can be instituted by any senior clinician with training in counselling for genetic testing.
Predictive testing is where there is a known genetic cause of a condition in the family, and you are testing to see if the patient is at risk of it. Currently only clinical genetics does this. The counselling is different. If someone asks you to do genetic testing in a predictive context, you can store DNA, but refer the baby / child to clinical genetics. They can link up the patient with the specific genetic variant to test only for that.

There are some conditions e.g. a newborn in a family with long QT where you might want to know the result relatively quickly. In these circumstances it is very reasonable to refer to the clinical genetics on call team (029218 42577 or se.genetics@wales.nhs.uk) to ask for expedited predictive counselling. Store DNA as above.

WINGS testing
If a baby / child is in NICU (neonatal intensive care unit) or PICU (paediatric intensive care unit), then WINGS (Wales Infants' and childreN's Genome Service) may be in an option if there are two unaffected parents available. This is rapid trio WGS (whole genome sequencing). NICU and PICU staff counsel and seek consent for it in conjunction with the on call clinical genetics team. Discuss with NICU / PICU staff if you think this is relevant and they have not raised it.

Routine trio WGS is also available only through clinical genetics.

If in doubt, discuss with seniors / refer as appropriate.

Contributors: Francis Sansbury, Jenny Gardner, Dirk Wilson

5.2 Electrocardiogram ECG in paediatrics

During working hours these are requested via the Cardiac Physiology Department (43198). This applies to inpatient and outpatient requests. Fill in a yellow request form.

ECGs may be performed out-of-hours using the machine kept on Pelican Ward. Familiarise yourself with the use of this machine.

Patients admitted with a suspected arrhythmia should have a 12-lead ECG in the arrhythmia, during pharmacological or other cardioversion therapy, and once sinus rhythm has been restored.

5.2.1 Basic ECG Interpretation

For more detailed information about quantitative ECG interpretation (e.g. age-related intervals), look on the “Measurements” section of the shared directory (S:\PaedCard\shared\MEASUREMENTS).
When assessing and ECG you should determine the following:

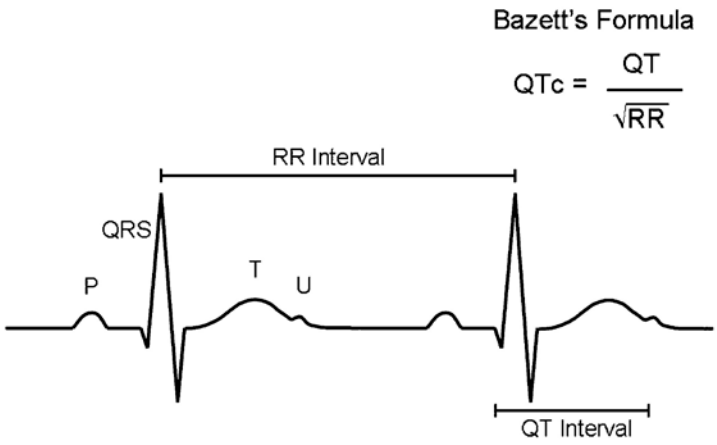
- Correct patient demographics
- Heart rate
- Rhythm
- Electrical axis
- PQRST morphology
- Ventricular forces
- Intervals

- PR
- QRS duration
- QT

The ECG computer package will calculate the intervals for you, but errors in the QT measurement are common and you should ideally calculate the corrected QT interval (QTc) yourself. A QT interval >440-460 msec could signify the presence of long QT syndrome where the use of certain drugs is contraindicated (for a list please see https://crediblemeds.org/ or download the app).

The most commonly-used method for QTc calculation is the Bazett formula:

QTc = QT/√RR interval



The Bazett formula over-corrects for slow and under-corrects for fast heart rates.

An alternative method, particularly for faster heart rates (>100 bpm), is the Framingham formula:

QTc (in msec) = QT (in msec) + (1000 – RR interval in msec / 6.5)

5.3 Exercise Test

Most children over 6 years of age should be able to manage the exercise treadmill.
Contact the Cardiac Physiology Department and fill in a request form. An outpatient letter or clinical summary should accompany the request. Use the paediatric request form and specify whether the test is cardiac physiologist or physician led.

Exercise testing allows staged increase in workload whilst measuring physiological parameters

- HR and BP
- Heart rhythm
- ST segments
- Workload (in metabolic equivalents, or METS)
- With cardiopulmonary exercise testing – peak oxygen consumption (VO_2), saturation data and respiratory function

Other measurements employed on occasion:

- Saturation data on standard exercise test (does the patient desaturate on exertion?)
- Peak expiratory flow rate pre- and post-exertion (is there exercise-induced asthma?)
- Echocardiography (how high does the aortic gradient go on exertion?)

5.3.1 Indications for exercise test

These include:

- Objective assessment of exercise tolerance
- Evaluation of exercise-induced symptoms, e.g. chest pain, dizziness, syncope

- Assessment of sinus node disease
- Provocation of exercise-induced arrhythmias
- Heart rate response in complete heart block
- Risk assessment in WPW
- Assessment of long QT syndrome (**use modified Bruce protocol, regardless of age – see section 5.3.8**)
- Assessment of peak oxygen consumption (VO_2 peak) and aerobic threshold (cardio-pulmonary exercise test, CPET, or CPEx)
- Assessment of severity of asymptomatic severe aortic stenosis*
- Risk assessment in hypertrophic cardiomyopathy*

*meticulous attention to BP required (see below)

5.3.2 Bruce protocols

The usual method of assessment at UHW is the exercise treadmill. It provides information on the haemodynamic and electrocardiographic response to a staged, progressive increase in load over a short period of time. In relatively fit, older patients the **standard Bruce protocol** should be used (arrange with the cardiac physiologist).

Standard Bruce stages:

Stage	Minutes	% grade	km/h	MPH	METS
1	3	10	2.7	1.7	5
2	6	12	4.0	2.5	7
3	9	14	5.4	3.4	10
4	12	16	6.7	4.2	13
5	15	18	8.0	5.0	15
6	18	20	8.8	5.5	18
7	21	22	9.6	6.0	20

Modified Bruce:

- Stage 1: 1.7 mph and 0% grade
- Stage 2: 1.7 mph and 5% grade
- Stage 3 corresponds to the first stage of the standard Bruce protocol
- Used in
  - Younger patients
  - Patients with moderate or severe exercise limitation
  - Patients being assess for long QT syndrome (allows careful calculation of QT interval with gentle changes in heart rate)

A bicycle ergometer is available for patients with mobility problems or who cannot manage the treadmill.

Exercise tests in low risk patients are now performed by the cardiac physiology team. High risk patients are overseen jointly with a doctor. High risk tests include those in patients with:

- Exercise-induced syncope of unknown cause
- Provocation of ventricular arrhythmia
- Severe aortic stenosis
- Cardiomyopathy
- Pulmonary hypertension
- A pacemaker
- A history of exercise-induced syncope

- Long QT syndrome, WPW, or a suspicion of exercise-induced arrhythmia
- A known coronary problems, e.g. ALCAPA, or Kawasaki disease and coronary artery aneurysm

Exercise test protocol:

- Check resting ECG (physiologist) and BP (doctor)
- Checking standing ECG (physiologist)
- Patient commences exercise
- Check BP and 12-lead ECG every 3 minutes (prompted by machine)
- Watch for arrhythmias
- Continue until either
    - The patient is exhausted
    - There is a medical indication to stop
    - The patient has reached the end of Stage 5 (standard Bruce), or
    - The target HR (THR) is achieved (i.e. 220 minus patient's age – we like to see a minimum of 85-90% of THR to show a "good effort")
- Document reason for cessation
- Take report to consultant's secretary (a report will be entered on Cardiobase by the consultant or one of the registrars); an electronic copy of the test will also be available on MUSE
- **If the test was terminated early because of an unexpected abnormality, please discuss this with the patient's consultant, or the service consultant before the patient is allowed home**
- If a doctor is helping to supervise a CPEx test, the role of the doctor is similar to that described above, although the physiologist will have a more complex role.

5.3.3 Medical indications for termination:

- Genuine patient exhaustion
- Significant arrhythmia (SVT, increasing ventricular ectopy, VT, complete heart block)
- Hypotension or failure to increase BP with symptoms of dizziness or chest pain; hypertension with systolic BP >220-240
- Severe chest pain with particularly with ECG changes (see below)
- Significant ST depression (flat or downsloping ST depression or ST elevation >2-2.5 mm in the absence of pre-existing bundle branch block) with or without symptoms

5.3.4 Expected responses:

- Ideally, achieve THR (220 minus patient's age)
- Achieve at least 85-90% of predicted peak heart rate
- Appropriate rise in systolic BP
- Minimal change in diastolic BP
- Expectation of exercising at least to the end of stage 4
- Absence of chest pain or excessive breathlessness
- Absence of arrhythmia
- No significant change in ST segments
- Recovery to baseline HR and BP within 7 minutes.

Normal responses in Welsh children (courtesy of Prof Orhan Uzun):

Age	9 to 10 years	11 to 12 years	13 to 14 years	15 to 16 years
No of subjects	All (n=22)	All (n=36)	All (n=46)	All (n=33)
Exercise duration (min)	13.1 ± 0.67	13.3 ± 0.50	13.1 ± 0.33	14.3 ± 0.49
Exercise capacity (METS)	14.7 ± 0.61	15.5 ± 0.45	15.4 ± 0.41	16.5 ± 0.51
VO_2 max ml/kg/min	51.6 ± 2.1	54.4 ± 1.6	53.7 ± 1.4	57.8 ± 1.8
Recovery time (min)	4.8 ± 0.30	6.3 ± 0.34	6.2 ± 0.24	5.7 ± 0.35
HR max (bpm)	194.3 ± 2.8	195.2 ± 2.0	196.4 ± 2.4	198.6 ± 2.2
% HR max reached (%)	92.3 ± 1.3	93.6 ± 0.99	95.1 ±1.2	97.1 ± 1.1
SBP max (mmHg)	136.1 ± 3.5	139.8 ± 2.0	146.5 ± 2.4	151.5 ± 2.9
DBP max (mmHg)	70.5 ± 2.1	70.9 ± 1.3	72.8 ± 1.3	73.5 ± 1.6
Rate pressure product (HRmax*SBPmax)	26567.1 ± 910.1	27275.8 ± 483.4	28824.9 ± 634.2	30125.7 ± 725.9

Estimated VO_2 max = METS x 3.5

5.3.5 Exercise test report

- Enter the report on Cardiobase
- Document resting ECG/HR/BP
- State which protocol was used
- State the duration of exercise and the workload achieved – comment on patient motivation
- State the reason for stopping
- State if HR and BP responses were normal – did the patient achieve >85% of predicted heart rate – if not the test was probably sub-maximal
- Comment on any ECG changes (rhythm, ST segments, QT interval if that is the question)
- State whether the test was
    - Normal
    - Abnormal

5.3.6 Difficulties in interpretation

- Breathlessness due to other cause (e.g. asthma)
- Pre-existing BBB, especially LBBB
- LVH
- WPW
- Digoxin (affects ST segments – consider stopping drug prior to exercise test)
- β-blocker therapy (blunts HR response and may mask ischaemia– consider stopping drug prior to test)

5.3.7 Helpful tips

- Continual gentle encouragement of the patient will maximise the information obtained in the study – try not to ask “do you want to stop” unless you think the patient is approaching or in genuine distress
- BP should be measured with the patient’s arm lifted off the equipment and supported by the operator – this minimises machine noise
- If there are difficulties auscultating the BP, use the palpation technique (radial or brachial artery)
- If you are unconvinced by the BP during exercise, check it again immediately after the treadmill stops and before the patient sits down – this can be taken as a surrogate for peak BP
- Meticulous BP measurement is essential in patients being exercised with HCM – a flat BP response (failure of systolic blood pressure to rise by more than 20–30 mm Hg from baseline), drop in BP during exertion, or precipitate drop in BP post-exertion are all poor prognostic signs (*Circulation.* 1997;96:2987-2991, *J Am Coll Cardiol* 2000;**36**:2212–8)

- The need for careful BP measurement also applies in AS – a flat BP response signifies important AS

Contributors: Shane Exton, Orhan Uzun, Dirk Wilson

5.3.8 ETT for Long QT Syndrome, including Schwartz criteria

On the request form, specify "Paediatric LQTS Protocol". A modified Bruce test should be performed. QTc can be measured at each stage, including 1, 2 and 4 minutes post recovery.
If QT hysteresis is to be performed, see:
S:\PaedCard\SHARED\MEASUREMENTS\EXERCISE\LONG QT Exercise Protocol

Contributors: Orhan Uzun, Amos Wong

The Schwartz criteria for diagnosing LQTS are shown below:

		Points
Electrocardiographic findings *(in absence of acquired causes)*		
QTc duration *(calculated by Bazett formula)*	≥ 480 msec	3
	460-479 msec	2
	450-459 msec *(men)*	1
QTc 4th minute of recovery from exercise test	≥ 480 msec	1
Torsades de Pointes* *(mutually exclusive)*		2
T-wave alternans		1
Notched T wave in 3 leads		1
Low resting heart rate	Below the 2nd percentile for age	0.5
Clinical history		
Syncope* *(mutually exclusive)*	With stress	2
	Without stress	1
Congenital deafness		0.5
Family history *(the same family member cannot be counted twice for the rows below)*		
Family members with definite LQTS	Index →	1
Unexplained SCD <30 years among immediate family members	Index →	0.5

Diagnostic criteria for long QT syndrome (LQTS) (the 'Schwartz-score'). Definite LQTS is defined by an LQTS score ≥3.5 points, intermediate probability of LQTS by an LQTS score of <3.5 and >1 and a low probability of LQTS by ≤1 point. In the family history rows, the same family member cannot be counted in both categories. **From https://heart.bmj.com/content/108/5/332**

5.4 Tilt Test

Indications:

- Investigation of repeated unexplained collapse, where high risk markers for cardiac syncope have been excluded

- Suspicion of vasovagal syncope (a.k.a. neurocardiogenic syncope or neurally mediated syncope), but atypical symptoms
- Assessment prior to drug therapy
- Patient education or tilt training (not currently available due to limited slots)
- Assessment of suspected postural orthostatic tachycardia syndrome (POTS) – ideally do a standing test to rule out general orthostatic hypotension – see https://www.cmaj.ca/content/194/10/E378

Procedure:

- The test is booked through the Cardiac Physiology Department using the requisite form
- Most tests are overseen by a cardiac physiologist with PLS certification – high risk cases may be overseen by a doctor as well
- Check resuscitation equipment and drugs prior to beginning
- Ensure you have a drug chart and a supply of GTN (400 microgram dose)
- Explain to the patient and parents what the procedure involves; mention that if GTN is used it may cause a headache, metallic taste and/or palpitations
- No IV line is required for sublingual GTN Tilt Test but make sure that an immediate access to IV line insertion is available in any emergency situation.
- The cardiac physiologist will "wire up" the patient and secure the supporting straps on the tilt table
- Continuous heart rate and BP monitoring is required
- Allow the patient to lie supine for 10-15 mins

- **Stage I: Drug Free State:**
    - Tilt the table (head up!) to 70 degrees for 20 minutes, recording heart rate and BP continuously
    - The person overseeing the test makes a note of any significant symptoms (nausea, sweating, presyncope) and relates these to physiological parameters.
    - If the patient blacks out lay the table flat immediately and ensure the ECG is being recorded continuously
    - If the patient does not experience syncope within 20 minutes, **proceed directly to Stage II without returning the patient to supine**

- **Stage II: Pharmacological challenge with sublingual GTN 400-500 microgram spray:**
    - Administer sublingual GTN and continue the tilt for a further 15-20 minutes.
    - Make a note of any significant symptoms (nausea, sweating, presyncope) and note the BP
    - If the patient blacks out lay the table flat immediately and ensure the ECG and blood pressure are being recorded continuously
    - If no syncope occurs with GTN terminate the procedure and return patient to supine position. Allow patient to rest for 10-20 minutes until they feel ready to stand up.
- Provide a written assessment of the test on the print-out. Discuss the results with the requesting consultant and ensure a report is entered into Cardiobase (found in a tab on the exercise test module)

Contributors: Shane Exton, Dirk Wilson

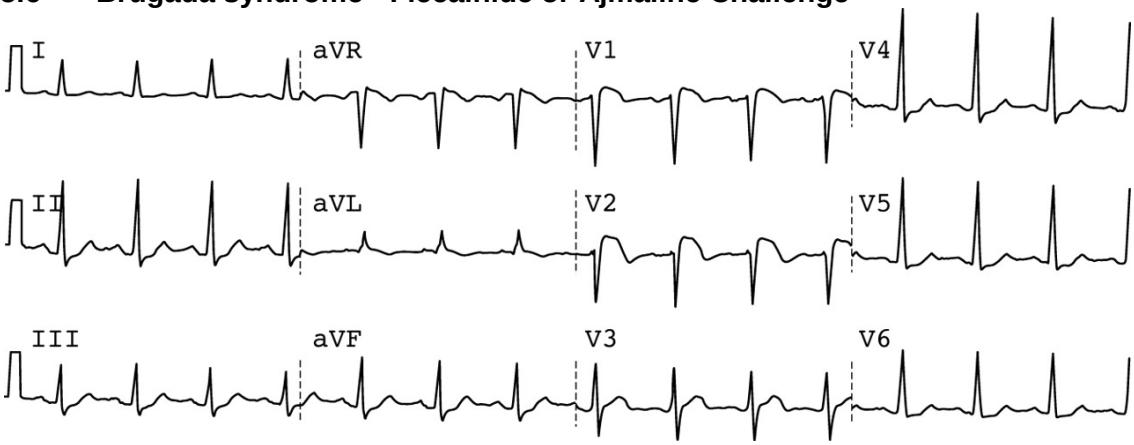
5.5 Adrenaline/Epinephrine challenge for LQTS

See shared directory: S:\PaedCard\SHARED\MEASUREMENTS\EXERCISE

Adrenaline challenge may be utilised, but genetic testing is more likely to yield diagnostic information.

Contributors: Orhan Uzun, Amos Wong

5.6 Brugada syndrome - Flecainide or Ajmaline Challenge



http://pmj.bmj.com/content/80/950/723/F2.large.jpg

- Brugada syndrome is an autosomal-dominantly inherited cause of sudden cardiac death and is generally due to mutations of the SCN5A gene on chromosome 3.
- The hallmark ECG features of RBBB pattern with ST elevation in leads V1-V3 are not always obvious in the resting ECG, but may be invoked or accentuated by blocking myocyte sodium channels using Class I anti-arrhythmic agents, e.g. flecainide or ajmaline.

Indications:

- Suspected Brugada syndrome (abnormal ECG with symptoms of palpitation or syncope)
- Screening of first degree relative with a confirmed diagnosis of Brugada syndrome (usually with symptoms or suspicious ECG).

Consider referral to the Bristol Electrophysiology Team for this procedure to be performed. If the decision is taken to perform the test locally, a ward bed must be booked in advance, ideally 2 weeks before – there must be the requisite nursing and junior doctor support and the resuscitation trolley must be immediately to hand. Inform ECG department well in advance of the procedure.

For full Ajamline / Flecainde Challenge protocol, see “Supplementary Information” section in the Wardbook folder in the shared directory:
S:\PaedCard\SHARED\WARDBOOK\FROM 2023\Supplementary Information

5.7 Six Minute Walk Test

The 6 minute walk test (6MWT) is the distance a person can walk at a constant, uninterrupted, unhurried pace in 6 minutes. It is being used increasingly in assessing the functional status of patients with severe cardiovascular disease, such as pulmonary arterial hypertension and heart failure. Its value is that, if repeated, it can provide longitudinal data

The test is either undertaken by the Respiratory Lab in Llandough Hospital – you may need to have a discussion with the lab about patients with pre-existing desaturation – or the paediatric cardiac nurse specialists. The following parameters are measured:

Resting state	Heart rate Saturations Perceived work of breathing (Borg index – see below)
During exercise	Heart rate Saturations Borg index

Distance walked at own pace

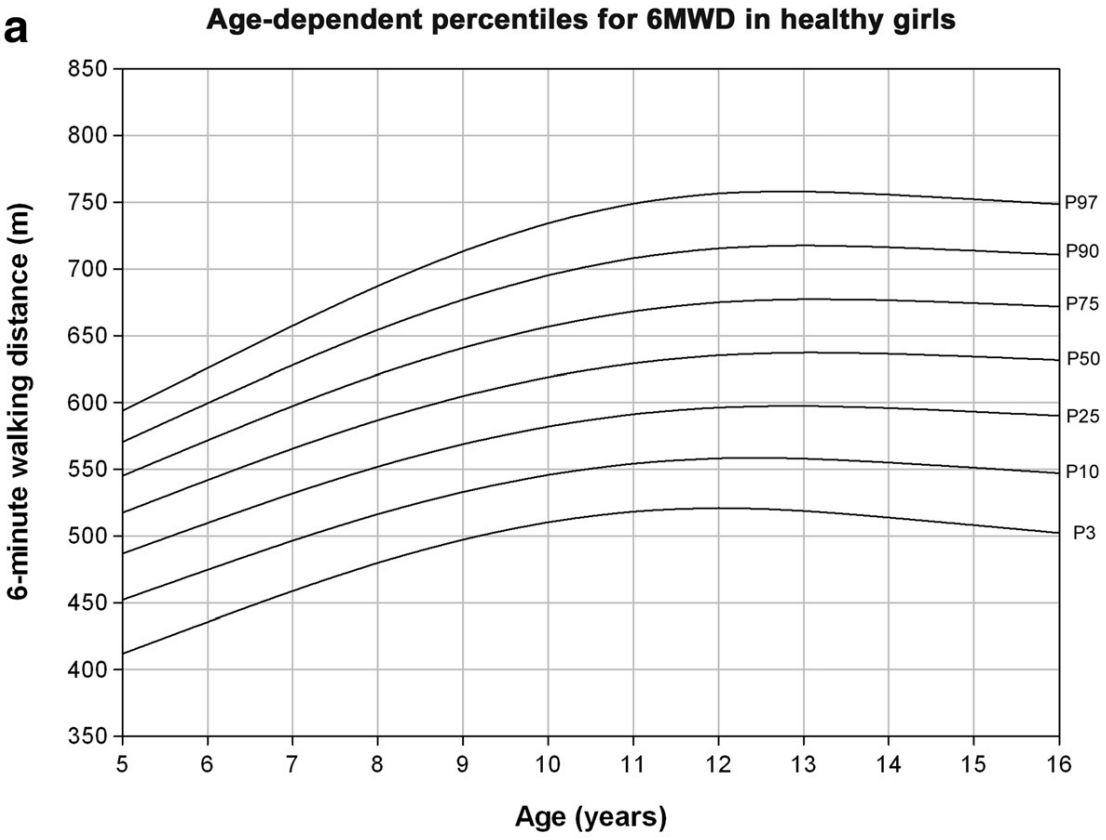
After exercise — Heart rate
Saturations
Borg index

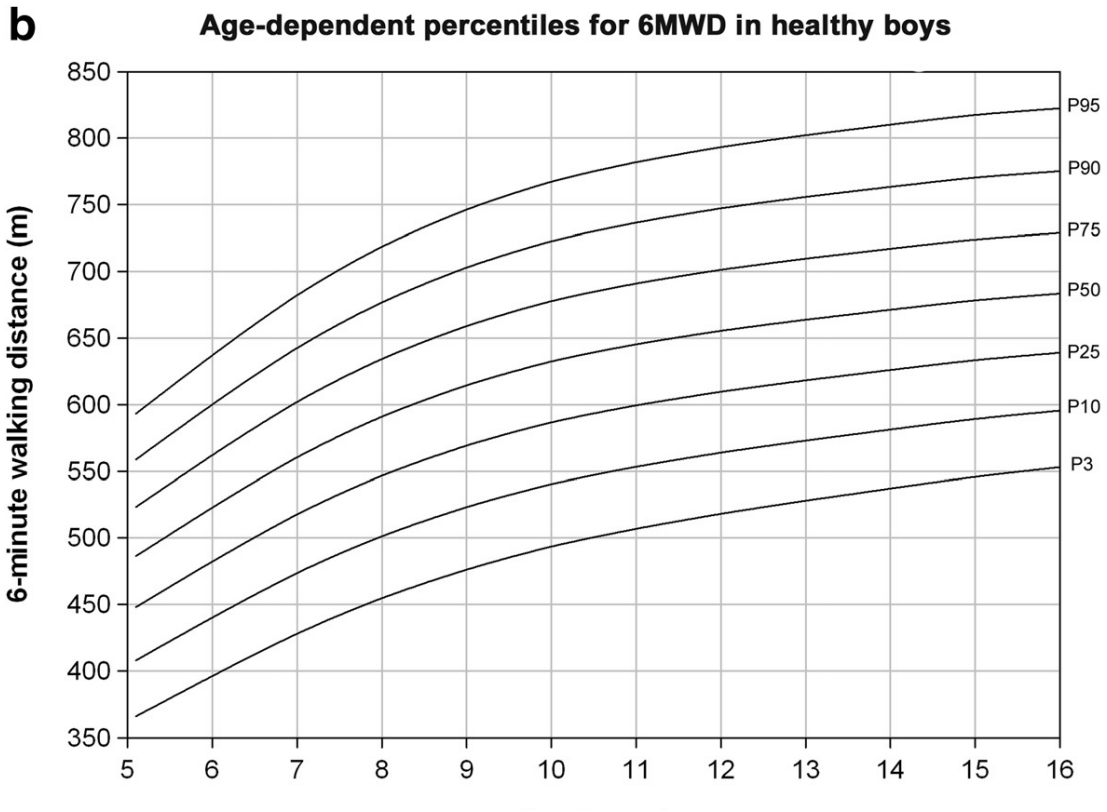
The modified Borg index is a vertical scale quantified from 0 to 10, in which 0 represents a patient's perception of no symptoms and 10 represents the perception of maximum breathlessness. It therefore provides and individual measurement of the intensity of the exercise.
The scale is derived as follows (based on patient/parent perception of breathlessness):

Borg index score	Perceived level of breathlessness
0	Not breathless
0.5	Very, very slight – breathlessness is just noticeable
1	Very slight
2	Slight
3	Moderate
4	Somewhat severe
5	Severe
7	Very severe
9	Very, very severe – just below maximum
10	Maximum – the most breathlessness one could endure

Values of 6 and 8 are not ascribed.

See overleaf for 6MWT normal values (https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3750631/)





References: Arch Dis Child 2008;93:464–468, **Contributors: Wendy Williams, Dirk Wilson**

5.8 Ambulatory Monitoring

This is requested through the ECG Department (46396). Reports should not be filed in the patient record until the result has been entered on to Cardiobase.

5.8.1 Holter monitor

Uses include:

- Investigation of palpitations, atypical seizures and syncope of unknown aetiology
- Assessment of severity of known arrhythmias
- Assessing cardiac rhythm in those at risk of arrhythmia (e.g. tetralogy of Fallot, post Senning or Mustard procedure, Fontan circulation, pre-excitation on ECG, 1st or 2nd degree heart block)
- Risk assessment of cardiomyopathy, CHB and LQTS
- Assessment of pacemaker function
- Assessment of response to anti-arrhythmic medications

Holter monitoring can be undertaken in any age group and can run from 24 hours up to one week (you need to specify the required duration on the request).

5.8.2 Holter reporting

Elements expected in a 24 hour tape (Holter) report on Cardiobase:

- Baseline demographics
- Reason for test
- Date and duration of recording
- Minimum HR
- Mean HR
- Peak HR
- A-V Conduction
    - ?Normal/Abnormal

    - ?Non-conducted or missed beats
    - ?Longest pause (significance)
    - ?Evidence of A-V block (timing, e.g. during sleep)
- SVE activity (%)
- VE activity (%)
- Tachyarrhythmia Y/N (description)
- Conclusion/Recommendations

Ensure the report is verified by a consultant and that it is filed in the patient record. The key findings should be summarised in a letter to the GP with cc's to the parents and local DGH.
Contributors: Shane Exton, Orhan Uzun, Dirk Wilson

5.8.3 Event recorders

The type of portable event recorder used at UHW is the R-Test. They are issued for up to two weeks. The device can be set up as a loop recorder (attached via leads to the chest continuously; when the patient presses a button the device saves the latest loop) or placed on the chest for recording during symptoms (patient activated). Rarely, a recording device may be implanted subcutaneously (Reveal device or Linq device), but this is reserved for severe, infrequent episodes. In patients with a Reveal if an episode is experienced, the patient needs to attend the Physiology Department for the device to be interrogated. Linq devices are uploaded to the cloud (the family will have instructions from the implanting centre).

The department has purchased, from charity monies, several patient activated recording devices, including the Alivecor Kardia device and the Withings Move ECG watch. Speak to the CNS team if there is a patient in whom such a device is needed.

5.9 Ambulatory Blood Pressure Monitoring

Blood pressure varies significantly over a 24 hour period and single, elevated measurements do not necessarily reflect the true situation. Ambulatory monitoring during routine daily activities provides several readings throughout the day and night, and offers a profile of blood pressure during rest as well as activities. It is especially important in differentiating spuriously high readings (white coat effect) from true sustained hypertension. It is helpful in:

- Assessing whether anti-hypertensive therapy should be commenced (e.g. after CoA repair)
- Assessing BP control in patients taking anti-hypertensives
- Deciding on optimal time to take anti-hypertensive medications
- Evaluation of hypotensive symptoms in patients taking anti-hypertensives, diuretics and ACE inhibitors

The cuff is permanently on the arm and the recorder placed on a belt or rucksack. The non-dominant arm is normally used, but in **coarctation patients it is mandatory to check the right arm BP** (there may be a BP gradient between the upper limbs).

There are several ambulatory monitors available for use in paediatric patients. Most use the oscillometric technique and correlate the reading with simultaneous recording of the heart rate which is also plotted on the graph. These monitors print the actual readings and mean BP values during time periods throughout the 24 hours.

Treatment decisions should be based on the mean values over the monitored period and take into account the shape of the profile. Normal 24 hour profiles show a fall in the blood pressure level during sleep - evidence suggests that patients with no fall (non-dippers) have an increased risk of end organ damage. Oscillometric mean ambulatory BP values in healthy children are shown below

by age and by height.

	Boys						Girls					
	Day			Night			Day			Night		
Age (years)	75th	90th	95th	75th	90th	95th	75th	90th	95th	75th	90th	95th
5	116/76	120/79	123/81	99/59	103/62	106/65	114/77	118/80	121/82	100/61	105/66	108/69
6	116/76	121/79	124/81	100/59	105/63	108/66	115/77	120/80	122/82	101/61	106/65	110/68
7	117/76	122/80	125/82	101/60	106/64	110/67	116/77	121/80	123/82	102/60	107/65	111/67
8	117/76	122/80	125/82	102/60	108/64	111/67	117/76	122/80	124/82	103/60	108/64	112/67
9	118/76	123/80	126/82	103/60	109/64	112/67	118/76	122/80	125/82	103/59	109/64	112/67
10	119/76	124/80	127/82	104/60	110/64	113/67	119/76	123/79	126/81	104/59	110/64	113/67
11	121/76	126/80	129/82	105/60	111/64	115/67	120/76	124/79	127/81	105/59	110/63	114/66
12	123/76	128/80	132/82	107/60	113/64	116/67	121/76	125/80	128/82	105/59	110/63	114/66
13	126/76	131/80	135/82	109/60	115/64	119/67	122/77	126/80	129/82	106/59	111/63	114/66
14	129/77	134/80	138/82	112/61	118/64	121/67	123/77	127/80	130/82	106/59	111/63	114/65
15	132/77	137/81	141/83	114/61	120/64	123/66	124/77	128/80	130/82	107/59	111/63	114/65
16	135/78	140/81	144/84	117/61	123/64	126/66	124/77	129/80	131/82	107/59	111/63	114/65

The values are in mmHg. Data from [46].

	Boys						Girls					
	Day			Night			Day			Night		
Height (cm)	75th	90th	95th	75th	90th	95th	75th	90th	95th	75th	90th	95th
120	116/77	122/80	125/82	99/58	103/61	106/63	114/77	118/80	120/82	99/60	103/63	106/65
125	117/76	122/80	125/82	100/58	105/61	108/63	115/77	119/80	121/82	100/60	104/63	107/66
130	117/76	122/80	126/82	101/59	106/62	110/64	116/76	120/80	122/82	101/59	106/63	108/66
135	117/76	123/80	126/82	102/59	108/63	111/65	116/76	120/80	123/82	102/59	107/63	109/66
140	118/76	123/80	126/82	104/60	109/63	113/65	117/76	121/80	124/82	103/59	108/63	110/66
145	119/76	124/79	127/81	105/60	111/64	114/66	118/76	123/80	125/82	103/59	109/63	112/66
150	120/76	125/79	128/81	106/60	112/64	116/66	119/76	124/80	127/82	104/59	110/63	113/66
155	122/76	127/79	130/81	107/60	113/64	117/66	121/76	125/80	128/82	106/59	111/63	114/66
160	124/76	129/79	133/81	108/60	114/64	118/66	122/76	126/80	129/82	106/59	111/63	114/66
165	126/76	132/80	135/82	110/60	116/64	119/66	123/77	127/80	130/82	107/59	112/63	114/66
170	128/77	134/80	138/82	112/61	117/64	121/66	124/77	128/80	131/82	108/61	112/67	115/71
175	130/77	136/81	140/83	113/61	119/64	122/66	125/78	129/81	131/82	109/59	113/63	115/66
180	132/77	138/81	142/83	115/61	120/64	124/66	N/A	N/A	N/A	N/A	N/A	N/A
185	134/78	140/81	144/84	116/61	122/64	125/66	N/A	N/A	N/A	N/A	N/A	N/A

The values are in mmHg. N/A, not available. Data from [46].

Contributor: Dirk Wilson

Reference: Lurbe: J Hypertens, Volume 27(9).September 2009.1719-1742

5.10 Echocardiogram

Please refer to https://pedecho.org/ for more background on echo, its standard views and moving images of paediatric / congenital echos (registration is required).

For more detailed information about normal values (including Z-score graphs), look on "Measurements" section of the shared directory (S:\PaedCard\shared\MEASUREMENTS) or use smartphone apps, e.g. Ped-Z (free) or Cardio Z (purchase).

During working hours, echos on cardiac patients are arranged via the Echo Physiologist (Paediatric Lead is Claire Roberts – Extension 43920, or use WhatsApp to contact her). Requests for echo should be communicated to the Physiology team early in the day. Their working hours are 0800-1600.

Echo referrals from other teams:

- There is almost no such thing as a referral for "echo only".
- Exceptions are follow-up neonatal echos (check known PDA), or chemo patients having LV function assessment between courses (if there have been no previous problems).
- All other echo requests are regarded as a "referral for cardiology opinion".
- Ensure the referral has been discussed by the child's lead consultant. Ensure the formal referral request document is filled in with adequate information.

- Remember – all echo reports are confirmed by a consultant - ensure echos for review are brought to the attention of the relevant consultant well before the end of the normal working day.
- If there are referrals for echocardiography out of normal working hours, this must be discussed with the consultant on call.

5.10.1 Echocardiogram under sedation

See C&V Paediatric Sedation Guideline:
General Paediatrics Clinical Portal - sedation-guideline-2013-currently-used-in-gen-paediatrics.pdf - All Documents (sharepoint.com)

It is common for infants to be admitted for echo under sedation. Admissions are arranged in advance through the medical secretaries. The degree of sedation required for successful echocardiography is "moderate" (refer to NICE guideline). **Print and use the Pro Forma for Paediatric Safe Sedation** found on the shared directory (next to the electronic version of this guideline book).
Before sedation: Ascertain the following:

- Patient weight and baseline observations
- Fitness of the patient for sedation
    - Patients who do not have significant respiratory disease or and airway problem should be NBM for ≥2 hours pre-sedation
    - In patients with significant respiratory disease or symptoms of airway obstruction the "fasting 2,4,6" rule should be applied (NBM 2^{o} for clear fluid (i.e. water), 4^{o} for breast milk and 6^{o} for formula milk or solids)
- Possible role of play therapist for toddlers (using distraction as an alternative to sedation)
- Possible need for and timing of ECG and CXR
- Availability of echo machine
- Availability of consultant to perform or complete the echo
- Obtain informed consent using a standard hospital consent form
- Document all clinical findings

Exposure: Ensure the top half of the patient is adequately exposed before the patient is asleep.

Monitoring: Saturation and ECG monitoring is required during and after sedation, until the patient is fully awake.

Sedation: When you have ensured the above, ascertain the consultant's preference, then the following advice should be followed:

Patient status	Drug and dose	Comment
< 6 months or <5 kg	Chloral hydrate 50 mg/kg po	Additional 25 mg/kg can be given if adequate sedation note achieved within 30 min
5-15 kg*	Chloral hydrate 75 mg/kg po	Maximum dose 1 g
>15 kg	Midazolam 0.5 mg/kg po Or Midazolam 0.2-0.3 mg/kg buccal	Maximum oral dose 20 mg <10 years, maximum buccal dose 5 mg ≥10 years, maximum buccal dose 7 mg

*If the child is very uncooperative from the outset, consider using oral midazolam, 0.5 mg/kg po, max dose 20 mg.

If the patient is disinhibited or restless after having the sedative, this may represent a paradoxical response – do not give further sedation. AVOID multiple sedative agents.

If midazolam is to be used, ensure ward availability of Flumazenil, in case of over-sedation/respiratory depression. See BNFc for dose and administration advice.

Inform the echocardiographer **as soon as the patient is sedated**, as the effects may only last a few minutes.

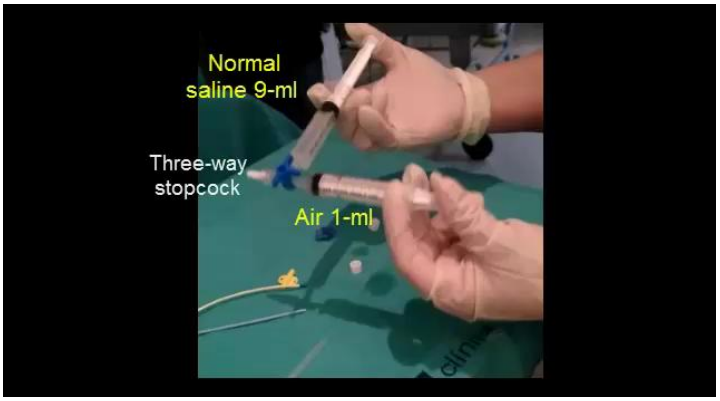
Discharge: Use the "safe sedation pro forma" to guide as to when the patient is fit for discharge. The discharging doctor should be satisfied that:

- Vital signs have returned to normal and that the airway, breathing and haemodynamic state have returned to baseline
- The patient is easily roused
- The patient has taken a feed

Document any problems in the sedation proforma, e.g. difficulties with sedation, and any complications, then the form should be filed in the patient record. When the patient is discharged, ensure the DAL is completed on Welsh Clinical Portal and a very brief summary is dictated.
Reference: www.nice.org.uk/guidance/CG112
Contributors: Gareth Davies, Chris Gillett and Dirk Wilson

5.10.2 Echocardiogram to rule out cardiac source of embolism ("bubble study")

Young patients with stroke should have a detailed echo to exclude a cardiac source of embolism. This includes assessment of right-to-left shunt across PFO with bubble-contrast echocardiography which requires an experienced echocardiographer and assistant. The recommended protocol is as follows:



- Intravenous cannulation is usually performed in the upper limb.
- The procedure for Valsalva with release is explained to both the subject and the parents. The subject is instructed to inhale deeply and exhale through the nose. Concurrently either the subject or the parent blocks the nasal passages externally with release on indication from the echocardiographer.
- Baseline transthoracic echocardiogram is performed. 5 ml of normal saline is drawn up into a 10 ml Luer-lock syringe and attached to a three-way tap with a further 10 ml Luer-lock syringe attached to another port. The final port is connected to the subject.
- The saline is agitated with air and the subject's blood (5 ml 0.9% saline to 0.5 ml of air and 0.5 ml of patient's own blood) by plunging it between the two syringes, ensuring that they are fully screwed on to prevent leakage of blood – see diagram below.
- When the saline mix is sufficiently agitated, initial injection with the patient breathing normally is performed to assess spontaneous passage of contrast into the left atrium. Sniffing can be requested at this time as this can sometimes open up a PFO and trigger a bubble shunt. A short, sharp sniff is needed.
- If no shunt is seen, the subject is instructed to exhale forcefully against the blocked nasal passages and mouth while a slow injection of agitated saline is performed.
- A slow injection is needed to ensure adequate contrast is present in the right atrium throughout the Valsalva. After an adequate Valsalva, where the left heart should become visibly smaller,

and with contrast present in the right atrium, the echocardiographer indicates to release the Valsalva and the number of bubbles crossing to the left atrium is assessed. This is repeated on up to five subsequent occasions, ensuring an adequate study. Adequacy in this case means that a Valsalva caused a reduced size in the left heart and was released when bubbles were filling the right atrium.

- Small shunt - <6 bubbles
- Medium shunt 6–20 bubbles
- Large shunt >20 bubbles
- Massive shunt - there would be dense microbubble opacification of the left ventricular blood pool.

False-positives:

- Pulmonary arteriovenous malformations (these always give a shunt without Valsalva, bubbles can be visualised coming down the pulmonary veins usually within 3 heart beats
- Use of colloid rather than saline to mix the contrast (in some patients, colloid contrast may pass through normal pulmonary capillaries allowing microbubbles to reach the left atrium when no PFO is present).

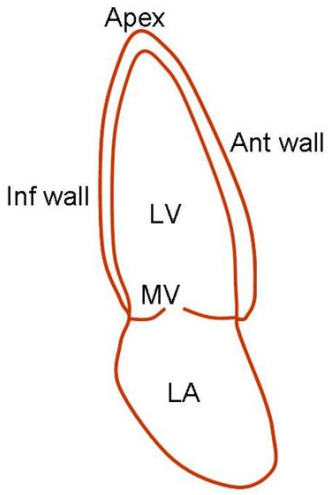
Reference: *Arch Dis Child 2008;93:255-259*
Contributor: Dirk Wilson

5.10.3 Echocardiogram for Ventricular Function

This is a common request from the NICU, PICU and Oncology services. Wherever possible, the following views should be obtained:

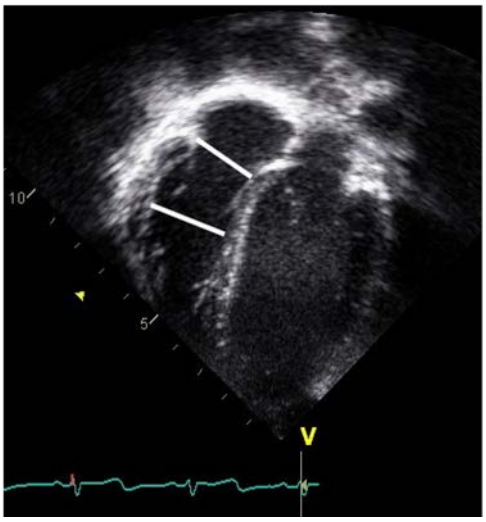
- Apical 4-chamber
  - MAPSE
  - TAPSE
  - Tissue Doppler of medial and lateral mitral annulus and lateral tricuspid annulus
  - Consider calculating ejection fraction using Simpson's technique
- Apical 2-chamber – This is obtained from the apical 4 chamber position. The probe is rotated counterclockwise 90°, so the notch will be around 12 to 1 o'clock. You should not see the right chambers. This view is used to assess the inferior and anterior walls of the LV. It gives a more precise evaluation of the direction and severity of mitral regurgitation.

https://web.stanford.edu/group/ccm_echocardio/cgi-bin/mediawiki/index.php/Apical_2_chamber_view

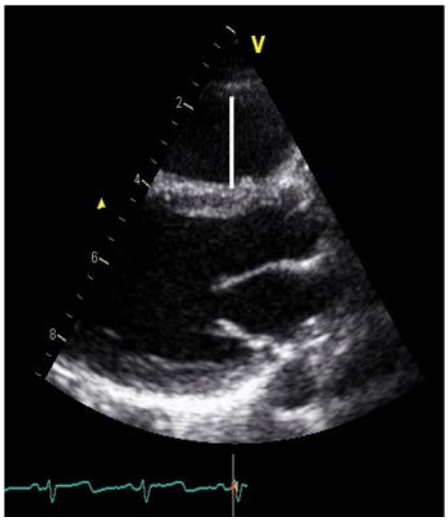


- Parasternal long axis
  - Fractional shortening

If RV function is in question, ensure the RV is measured in diastole in the 4-chamber (RV base and mid-RV) and long axis views (see figures below).



4-chamber RV measurements at base and mid-ventricle

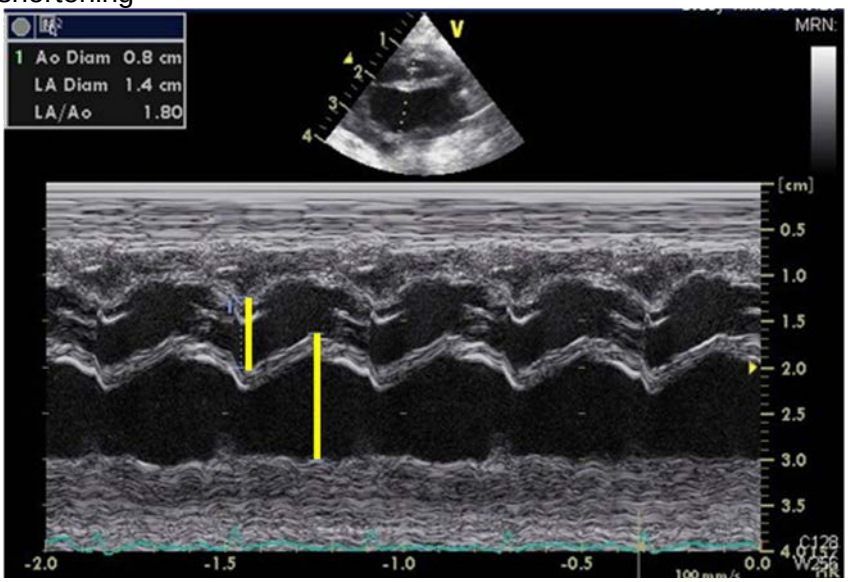


Long-axis RV measurement

Contributors: Orhan Uzun, Dirk Wilson, Amos Wong

5.10.4 Echo Assessment of PDA in Premature Neonates

1. Assess for presence of PFO / ASD
2. Assessment of left heart
    a. 2-D LA diameter in long axis view (mm)
    b. Left atrial area (cm^2 – trace in ventricular systole)
    c. LA to Ao ratio (Ao dimension on M-mode at early systole, LA dimension on M-mode at end ventricular systole, both leading edge to leading edge – see figure below)
    d. Left ventricular dimensions and function
    e. LV end systolic and end diastolic dimensions either on 2D or M-mode at level of tips of MV
    f. Using LV end diastolic and end systolic dimensions calculate fractional shortening



3. Assessment of PDA and its effects
    a. Record peak systolic arterial blood pressure at time of assessment
    b. Record whether or not there is a clear ductal ampulla
    c. Record minimum PDA size (mm) in short axis and “ductal view”
    d. Assess PDA using continuous and pulsed-wave Doppler (NB – ECG trace on)
    e. Record direction of PDA shunt (exclusive L→R, bidirectional, exclusive R→L)

f. Record highest systolic ductal velocity
g. Record highest diastolic ductal velocity
h. Record aortic flow using PW Doppler at level of diaphragm (subcostal view) – note whether there is diastolic flow reversal
i. Record superior mesenteric artery flow using PW Doppler – note whether there is absence of diastolic flow

4. Exclusion of other pathology
    a. Has the scan excluded duct-dependent congenital heart disease excluded?
    b. Has the scan excluded coarctation of the aorta or suspected arch pathology? Record the diameter of the aortic isthmus (segment of aorta between L subclavian artery and PDA).

Criteria that support the diagnosis of a haemodynamically significant duct:

- Minimum ductal dimension > 2 mm at its narrowest point
- LA:Ao ratio >1.5
- Peak left to right ductal maximum velocity <2m/s
- Holodiastolic flow reversal in the descending aorta at level of the diaphragm

Echocardiographic contraindications to PDA ligation:

- Duct-dependent congenital heart disease
- Coarctation of the aorta or suspected arch pathology
- Any right to left shunting at ductal level

Contributors: John Forsey, Orhan Uzun, Mark Walsh, Dirk Wilson

5.10.5 Transoesophageal Echocardiography

This is normally undertaken electively in Bristol. There may be emergency situations where a TOE is needed in Cardiff, e.g. trauma patient.

Consent: This is obtained by the person undertaking the procedure.

Please note: The paediatric probe is now always hung up in the cupboard in the TOE room located in the ECG Department, B1/ C1 Link corridor. After 5pm or if the department is locked ascertain if B1 has a key, and if not you will need to telephone Security to open up for you.

During working hours please ask a member of the adult echo physiologist team to prepare the probe for you. It has its own carrying tray and covers and will be sterilized for you but adequate warning (**several hours**) should be given to allow this to be completed in time. If you have to use it without being sterilised the probe must be thoroughly wiped over with a sterilising wipe and placed in the carrying tray.

Paediatric probe covers are still kept in the TOE room as before.

After use please carry probe back **in its tray** and inform the echo physiologists that it has been used and they will hopefully clean and resterilise it for us and replace it back into the cupboard. **Never replace it back into cupboard until it has been cleaned properly**.

Contributors: Claire Gemmell, Leann Gibbs, Dirk Wilson

5.11 Cross-Sectional Imaging – MRI and CT

- Useful in assessing cardiac anatomy (particularly the great vessels), cardiac function, severity of valve regurgitation and other haemodynamic issues.
- Scans are organised through the Consultant Cardiovascular Radiologist.
- Scans under GA have to be booked well in advance (see below).
- If the patient has pre-existing renal dysfunction and gadolinium contrast is to be used, there is a small risk of a rare but irreversible skin complication called nephrogenic fibrosing sclerosis. **In the cases where it is known gadolinium contrast will be used, to guard against this adverse effect:**

- It is now a requirement to provide the MRI coordinator with "up-to-date" U&E/creat (i.e. within 4 months of the scan).
- Depending on the age and location of the patient this test can be done by the GP, the local DGH or at UHW prior to the investigation.
- In practice it is best to have the results at least a week before the scan is due – try not to leave until day of admission for MRI.

5.11.1 Non-General Anaesthetic

Older patients will be sent an appointment by Radiology directly. Younger patients may have their scan via Pelican Ward, Owl Ward or the Children's Investigation Unit (CIU).

Admission: Children having cross-sectional imaging without GA should attend the ward 2 hours prior to the procedure.

IV Access: Venous access should be established as early as possible to enable a period of heart rate recovery (use Emla or Ametop).

Play Therapy: If the play therapist is to be involved, this should also be scheduled within this 2-hour window.

β-Blocker: Patients having **CT** and not already on a β-blocker should be prescribed propranolol 0.5 mg/kg PO 1 hour pre-procedure (maximum dose 40 mg) to attain a HR <80 bpm (ensure no contra-indication).

Consent: Obtained by clerking doctor.

State that iv contrast medium is likely to be administered [small risk of contrast reaction]; if for CT – dose of radiation carries a very small risk of tumour or leukaemia development (~1:2500 or less). There is also a risk of "unexpected findings" which may require further action.

Discharge: Patients who have undergone CT without GA may be discharged immediately after the procedure if there is no evidence of an anaphylactic reaction to the iv contrast medium.

5.11.2 General Anaesthetic

Admission: Children admitted for cross-sectional imaging under GA are routinely admitted the afternoon before the procedure to allow anaesthetic assessment.

IV access: Not necessary for MRI scan (line placed by anaesthetist)

For CT angiogram, there is a requirement for a heart rate < 80 bpm – this may be achieved by anaesthesia alone, but check with the consultant paediatric cardiologist / radiologist / anaesthetist whether propranolol should be prescribed (ensure no contraindication). If so, a dose of 0.5 mg/kg should be given 1-2 hours before the procedure. In infants, place an IV line and prescribe maintenance dextrose/saline for 4 hours pre-procedure to prevent hypoglycaemic reaction. If a β-blocker is not to be given pre-procedure, there is no need for line placement.

Consent: Obtained by clerking doctor (state that iv contrast medium is likely to be administered – small risk of contrast reaction); if for CT – dose of radiation carries a very small risk of tumour development (~1:2500 or less). Fast track patients will have been consented in the outpatient setting – check to see if this was done.

Discharge: Patients who have undergone CT under GA may be discharged when they are drinking normally and have managed something to eat. **NB – infants under 60 wks post conceptual age (i.e. 20 wks/5 months corrected age) must be kept in for observation overnight (small risk of apnoea).**

5.11.3 Surveillance post aortic stent implantation

Post CoA Stent – provided post-implant angiography was satisfactory:

Adults

- CT angiogram 3-4 months post implant.
- If satisfactory, continue 12-18 monthly echo surveillance
- Consider need for PA/lateral CXR (if concerns about stent position or integrity)
- Repeat CT angiography 5 yearly

Children

- CT angiogram 3-4 months post implant – referring clinician to judge whether this is GA or non-GA
- If satisfactory, continue 6-12 monthly echo surveillance
- Consider need for PA/lateral CXR (if concerns about stent position or integrity)
- If concerning features (evidence of stent fracture, hypertension, reducing femorals, increasing stent Doppler, increasing size of aorta, suspicion of aneurysm) discuss case with interventionist and refer for catheter assessment or CT angiogram
- Consider CT angiogram once growth is completed (unless there has been a recent catheter) and ~5 yearly thereafter

NB – post-implant CT angiogram is focused and confined from a few cm above to a few cm below the stent, unless there is concern about more widespread aortopathy.

Contributors: Rob Martin, Mark Turner, Dirk Wilson, Andrew Wood

5.11.4 Adenosine Stress Test by MRI

Refer to the "Supplementary Information" sub-folder in the Wardbook section on the Shared Directory S:\PaedCard\SHARED\WARDBOOK\FROM 2023\Supplementary Information

5.12 Isotope Scans

These use a small dose of radio-isotope, which is excreted in the urine. Ensure the child's mother is not pregnant! Sedation may be required; intravenous access will be required.

5.12.1 Cardiac Nuclear Scanning

Main indications include assessing myocardial ischaemia (myocardial perfusion imaging) and ventricular function. Requests need to be discussed with Dr Richard Wheeler (adult cardiologist).

5.12.2 Lung perfusion scan

Organise through Radiology. Indications are exclusion of pulmonary embolus (V/Q scan) and assessment of differential lung perfusion (e.g. in branch PS).

SECTION 6 – CLINICAL PROBLEMS

Section Index

6.1 Anti-platelet Therapy and Anticoagulation

6.1.1 Aspirin and Anti-platelet Therapy

Indications for use of aspirin in children (plus see tables below):

- Anti-inflammatory action – treatment of
    - Acute pericarditis
    - Kawasaki disease (acute phase)
    - Acute rheumatic fever

- Anti-platelet therapy – treatment of
    - Kawasaki disease (convalescent phase)
    - Systemic-pulmonary shunt
    - Chronic cyanosis (e.g. cavopulmonary shunt, Eisenmenger syndrome – relative indication)
    - Bioprosthetic valve (surgeon preference)
    - Prosthetic valve with history of embolism despite anticoagulation (added therapy)

If aspirin cannot be used (e.g. allergy), consider the use of other agents such as dipyridamole or clopidogrel (limited data in children).

In the event of development of chicken pox, herpes, influenza, rubella, or other severe flu-like febrile illness, the clinician will determine whether the risks / benefits of continuing aspirin vs the small risk of Reye's syndrome. Parents should be instructed to telephone to ask for advice in this situation. Patients on aspirin should not have Fluenz nasal immunisation – instead they should have the injected flu vaccine. Also consider varicella vaccination.

Patients on aspirin for a B-T shunt **should not have therapy discontinued**, even during a febrile illness; however patients with a weaker indication for aspirin (e.g. chronic cyanosis with a cavopulmonary shunt, Kawasaki disease) should discontinue aspirin temporarily during the feverish phase of an illness. The consultant may consider use of dipyridamole or clopidogrel during this period (NB the data sheet for clopidogrel also advises discontinuation during chicken pox, etc).

6.1.2 Devices, Patches and Stents

Indication	Treatment	Duration
ASD device	Aspirin 3-5 mg/kg once daily, usual maximum 75 mg once daily*	6 months
ASD surgical closure	Suture closure or patch closure with pericardium or Goretex – **no** antithrombotic therapy needed, unless other indication (e.g. supraventricular arrhythmia, pro-thrombotic condition, identified post-op thrombosis); Dacron patch closure – Aspirin 3-5 mg/kg once daily, usual maximum 75 mg once daily*	6 months
VSD device	Aspirin 3-5 mg/kg once daily, usual maximum 75 mg once daily*	6 months
Aortic stent	Aspirin 3-5 mg/kg once daily, usual maximum 75 mg once daily*	6 months
Branch pulmonary artery stent	Aspirin 3-5 mg/kg once daily, usual maximum 75 mg once daily*	6 months
PDA closure device	Not indicated	

PDA stent	Aspirin 5 mg/kg once daily plus Clopidogrel 0.25 mg/kg once daily	Until corrective surgery or Glenn
RVOT stent	Aspirin 5 mg/kg once daily	Until corrective surgery or Glenn
Sano shunt	Aspirin 5 mg/kg once daily PLUS Clopidogrel 0.2 mg/kg once daily	Until Glenn shunt

*NB – there may be individual clinical reasons to extend treatment, to supplement with other antiplatelet agents (e.g. clopidogrel), or to use formal anticoagulation. Aspirin dose of 150 mg or 300 mg may be used in older adolescents.
See section 6.1.1 for advice regarding use of aspirin during intercurrent illness.

6.1.3 Valve replacement

Indication	Treatment	Duration
Tissue valve (tricuspid, mitral, aortic)	Aspirin 3-5 mg/kg once daily, usual maximum 75 mg once daily*	6 months – longer if surgeon specifies; in some circumstances, the surgeon may specify 3-6 months of warfarin, followed by long-term aspirin
Tissue RV-PA conduit or pulmonary valve (any type, including transcatheter implant, Ross operation)	Aspirin 3-5 mg/kg once daily, usual maximum 75 mg once daily*	6 months – longer if surgeon/interventionist specifies; in some circumstances, the surgeon may specify 3-6 months of warfarin, followed by long-term aspirin
Prosthetic valve (any type including mitral or aortic)	Warfarin – target INR 3.0 ± 0.5 (i.e. range 2.5-3.5)**	Indefinite

*NB – there may be individual clinical reasons to extend treatment, to supplement with other antiplatelet agents (e.g. clopidogrel), or to use formal anticoagulation. Aspirin dose of 150 mg or 300 mg may be used in older adolescents.
See section 6.1.1 for advice regarding use of aspirin during intercurrent illness.
**NB

- In high-risk AVR or MVR patients (e.g. small valve or increased risk of thrombosis), a target INR of 3.5 may be used (range 3 – 4) – this needs to be clearly specified in the medical notes and in the INR booklet.
- Our unit experience tells us that an upper range of 4 has been used without adverse incidents in the past.
- AHA guidelines recommend a target INR of 2.5 (range 2.0 – 3.0) for aortic valve replacement with a Starr-Edwards valve or a tilting disk valve (other than Medtronic-Hall) with no other risk factors (reference *Circulation* 2013;128:2622-2703). Based on our local experience, this unit will continue to have a target INR of 3.0 (or higher) in all prosthetic paediatric aortic valves.

6.1.4 Cavopulmonary shunt / Fontan

Indication	Treatment	Duration
Glenn / superior cavopulmonary shunt	Aspirin 3-5 mg/kg once daily, usual maximum 75 mg once daily*	To continue until Fontan / TCPC**
Fontan / TCPC	Warfarin – target INR = 2.5 ± 0.5 (i.e. range 2-3) Consider use of Rivaroxaban or other DOAC in selected cases – see section 7.15	Indefinite***

*NB – See section 6.1.1 for advice regarding use of aspirin during intercurrent illness.
**NB – There may be individual clinical reasons to extend treatment, to supplement with other antiplatelet agents (e.g. clopidogrel), or to use formal anticoagulation.
***NB – Some patients may be treated with aspirin rather than warfarin – the reasons for this should be clearly stated in the surgical summary and the local patient record. AHA guidelines support the use of aspirin alone in uncomplicated Fontan patients, and Warfarin in patients with adverse risk factors (see *Circulation* 2013;128:2622-2703). Use of direct oral anticoagulant drugs (DOACs), such as Rivaroxaban or Apixaban, after Fontan has no evidence base, but may be considered in individual circumstances (e.g. older patient with difficult to control INR) following careful discussion with the family – see section 7.15.

6.1.5 Other indications

Indication	Treatment	Duration
ASD surgical patch with Dacron (pericardial / Goretex patch will not normally be given aspirin)	Aspirin 3-5 mg/kg once daily, maximum 75 mg daily	6 months
Kawasaki disease	Refer to AHA or UK guidelines	Refer to AHA or UK guidelines
Line related venous or arterial thrombosis	Clexane (infant) or warfarin (older child; target INR 2.5 (range 2-3)	3 months, with levels maintained in the treatment range
Modified Blalock-Taussig shunt	Aspirin 3-5 mg/kg once daily, usual maximum 75 mg once daily	To continue until next definitive surgery; treatment to continue with febrile illness
Pulmonary arterial hypertension (idiopathic, genetic or familial)	EITHER No antithrombotic therapy OR Aspirin 3-5 mg/kg once daily, usual maximum 75 mg once daily* OR Warfarin – target INR 2.0 (range 1.5-2.5)	Indefinite
Pulmonary arterial hypertension (Eisenmenger syndrome)	EITHER No antithrombotic therapy OR	Indefinite

	Aspirin 3-5 mg/kg once daily, usual maximum 75 mg once daily* OR Warfarin – target INR 1.8 (range 1.5-2.1) OR Rivaroxaban – see section 7.15	
Dilated cardiomyopathy	Aspirin 3-5 mg/kg once daily OR Warfarin – INR target 2.5 (range 2-3) OR Rivaroxaban – see section 7.15	Continue until fractional shortening >25%, and/or not deemed high risk of LV thrombosis
Post L heart electrophysiology ablation	Aspirin 3-5 mg/kg once daily, usual maximum 75 mg once daily*	3 months

*NB – there may be individual clinical reasons to extend treatment, to supplement with other antiplatelet agents (e.g. clopidogrel), or to use formal anticoagulation.
Aspirin dose of 150 mg or 300 mg may be used in older adolescents.
See section 6.1.1 for advice regarding use of aspirin during intercurrent illness.
References:

1. Guideline on antiplatelet and anticoagulation management in cardiac surgery *Eur J Cardiothorac Surg* 2008;34:73-92
2. Valvular and Structural Heart Disease. *Chest* 2008; 133: 593S - 629S.
3. Antithrombotic Therapy in Neonates and Children. *Chest* 2008;133;887S-968S
4. Prevention and treatment of thrombosis in pediatric and congenital heart disease: a scientific statement from the AHA. *Circulation* 2013;128:2622-2703

6.1.6 Commencing anticoagulation (heparin and warfarin)

Please refer to the all-Wales Paediatric Thrombosis and Anticoagulation Guideline:

Neonatology Clinical Portal - PAEDIATRIC THROMBOSIS AND ANTICOAGULATION GUIDELINES V7 NICU.pdf - All Documents (sharepoint.com)

Relevant parts of this guideline are reproduced below.

Low molecular weight heparin (LMWH)

Prior to therapy:

1. Exclude contraindications (see BNF for Children)
2. Measure full blood count and keep platelets >50,000, and check clotting screen
3. Measure renal function and be cautious in renal impairment (see below and discuss with Haematology)
4. Obtain blood group and cross match
5. Ensure adequate supply of blood products available for patients
6. Ensure adequate supply of protamine sulphate available
7. Perform cranial ultrasound scan in neonates

LMWH (enoxaparin): Neonate 1.5 – 2 mg/kg
1 - 2mo age: 1.5mg/kg
>2mo age: 1.0 mg/kg.

During therapy:
Phone laboratory to discuss timing of anti-Xa levels. Samples can be frozen and defrosted for a routine run. Prior to the assay the instrument (automated coagulation machine) must be purged/cleaned. This takes several hours and may mean the rest of the hospital will not be able to get coagulation results. Be careful what you call "urgent" results
Target anti-Xa is 0.5-1iu/ml
Administer 4 to 5 doses (2 to 3 days' worth) before checking levels.
Do NOT use insuflons or other S/C devices to administer doses – they cause marked variation in total drug dose delivered
Measure anti Xa 4 hours after dose and adjust according to table:

Anti Xa	Hold next dose	Dose change	Repeat Xa level?
<0.35U/ml	No	Increase by 25%	4h after 4 doses
0.35-0.49U/ml	No	Increase by 10%	4h after 4 doses
0.5-1.0U/ml	No	no	1 week later, then monthly while receiving enoxaparin (4h after am dose)
1.1-1.5U/ml	No	Decrease by 20%	Before next dose
1.6-2.0U/ml	3h	Decrease by 30%	Before next dose, then 4h after next
>2.0U/ml	Until Xa ≤ 0.5	Hold	Before next dose, If not ≤0.5, repeat q12h, then decrease dose by 40%

Levels are routinely run on Monday/Wednesday/Friday. Non-urgent samples can be frozen and analysed at a more convenient time. For urgent samples discuss with the on call coagulation team or paediatric haematology
Side effects and precautions: Bleeding

Use unfractionated heparin (UFH) in renal failure as LMWH is excreted by the kidneys. It may be possible to switch to LMWH once anticoagulation established – discuss with Haematology
Monitor for Thrombocytopenia. Check FBC on day 8 of therapy.
Avoid NSAIDS or anti-platelet drugs
Duration of therapy: Has been used for a short course 10-14 days. For extensive deep vein thrombosis heparin has been used for 3 – 6 months.

The child's carer will be competency assessed prior to discharge to ensure safe home administration of LMWH. Written instructions will be given to the carer on how to administer (see Appendix A for documents)

Unfractionated Heparin (UFH)

Prior to therapy:
1. Exclude contraindications (see BNF for Children)
2. Measure full blood count and keep platelets >50,000, and check clotting screen
3. Measure renal function and be cautious in renal impairment (see below and discuss with haematology)
4. Obtain blood group and cross match
5. Ensure adequate supply of blood products available for patients
6. Ensure adequate supply of protamine sulphate available on the unit
7. Perform cranial ultrasound scan in neonates

During Therapy

APTTR = patient's APTT / mid-point of normal range

For example, if the APTT is 32 seconds and the range is 26 to 38 seconds:

APTTR = 32/[(38-26)/2] + 26 = 32/(6 + 26) = 32/32 = 1

Maintain APTT between 60 and 85 seconds. This corresponds to an APTTR 1.5 to 2.5. Give by IV route

Loading dose: 75 U/kg over 10 minutes } Check with BNFc

Maintenance: 10 – 20 U/kg/hr (higher doses may be needed) }

Check APTT 6 – 8 hours after starting therapy.
If APTT ratio is LOW – increase maintenance dose by 10-20% and recheck APTT ratio 4-6 hours later.
If APTT ratio is >2 and ≤3, reduce the maintenance dose by 10% and recheck APTT ratio 6-8 hours later.
If APTT ratio is >3, stop heparin for 1 hour, then restart at a reduced maintenance dose (reduce by 20%).
NB – if maintenance doses of >35 U/kg/hour are required it may be appropriate to accept slightly lower APTT ratios.
If bleeding develops then stop the infusion and inform a senior. Consider protamine sulphate.
Administer protamine sulphate as follows (based on total amount of heparin received in last 2 hours):

Heparin (time since last dose, minutes)	**Protamine Sulphate Dose** (per 100 units of heparin received)
Less than 30	1 mg
30 – 60	500 – 750 micrograms
60 – 120	375 – 500 micrograms
Greater than 120	250 – 375 micrograms
Max Dose **Infusion rate**	**50 mg** **Infuse over 10 minutes** **(max rate 5 mg/minute)**

Warfarin
Procedure and process of prescription and parent competency assessment shall be done as per the All Wales Paediatric Warfarin Care Pathway and prescribed on the all Wales Paediatric In-Patient Warfarin Treatment Chart.

Copies of these documents are given in Appendix B of the policy (see C&V Web).

Tissue-type plasminogen activator (rt-PA)
Most often used in neonates to treat life or limb threatening thrombosis.

Paediatric Cardiology use it to treat limb ischaemia following femoral puncture (& thrombosis) for catheterization.

Also indicated in massive pulmonary embolism where there is systemic hypotension

Treatment of neonatal thrombosis is still controversial. The evidence based for the management of neonatal thrombosis is very limited and is mostly based on case series and extrapolated adult literature. Tissue type plasminogen activator (t-PA) has been used to treat both neonatal arterial and venous thrombosis.
Consider t-PA for the following Indications:

Any limb, life or organ threatening condition secondary to thrombosis.

Bilateral renal vein thrombosis with impending renal failure
Arterial thrombosis with impending loss of limb (femoral, iliac, axillary arterial thrombosis)
Extensive aortic or vena caval thrombosis
Intracardiac thrombosis compromising systemic or pulmonary circulation

Absolute contraindications for use:

1. Active bleeding at any site
2. Any General Surgery in the past 10 days or Neurosurgery in the last 3 weeks

Relative contraindications for use:

1. Thrombocytopenia (<50,000)
2. Low fibrinogen concentration (<100mg/dl)
3. Preterm <32 weeks

Prior to initiating therapy

1. Exclude contraindications
2. Ensure good venous access for drug administration and for monitoring purposes
3. Measure full blood count, fibrinogen
4. Obtain blood group and cross match
5. Notify blood bank to ensure FFP and cryoprecipitate are available.
6. Notify Pharmacy to ensure tranexamic acid is available
7. Perform cranial ultrasound scan in neonates
8. Ensure adequate venous access
9. Stop heparin infusion 3 hours prior to therapy
10. In Neonates give FFP 10-20ml/kg at least 30mts prior to starting thrombolytic therapy. (ACCP guidelines) to provide some plasmin for the drug to activate
11. In older children transfer to PICU or HDU prior to initiating therapy.

Dose Regimens -
There are no trial data to support one over the other.

Case series evidence favours the low dose regimen as less likely to cause severe bleeds

<u>High Dose:</u>
Give a loading dose of 0.1mg/kg, followed by an infusion of 0.3-0.5mg/kg/hr over 6 hours.

<u>Low dose:</u>
Give 0.1mg/kg/hr for 4 hours. Once the infusion is complete, start UFH (protocol as above) and continue until next rt-PA infusion.

Perform ultrasound scan of thrombosed vessel at the end of infusion and if recanalisation is not complete. Up to four additional doses of t-PA can be given at intervals of 12-24 hours.

During therapy:

1. No intramuscular injections during therapy

2. Minimal manipulation of the patient i.e. no bathing, physiotherapy
   Avoid concurrent use of coumadin or antiplatelet agents (i.e. NSAIDS, Aspirin, persantin).
3. No urinary catheterisation, rectal temperatures or arterial punctures
4. Blood samples from a superficial vein or indwelling catheter. If blood sampling is difficult, insert an indwelling catheter for blood samples **prior** to thrombolytic therapy
5. Monitor fibrinogen level 1 and 4 hour after each t-PA infusion. Expect a 20-50% drop in fibrinogen levels. Maintain fibrinogen level >1g/l with FFP or cryoprecipitate or fibrinogen concentrate infusion
6. Maintain platelet count >$100x10^9$/L
7. Do not give intramuscular injections and do not do procedures like urinary catheterisation, rectal temperatures, arterial punctures etc.
8. Minimal handling of patient
9. Perform daily cranial ultrasound scan
10. If a patient has received thrombolytic therapy for more than 6 hours, consider treating with heparin alone for 24 hours before reinstituting thrombolytic therapy. There may be ongoing thrombolysis even in the absence of continued administration of the thrombolytic agent

Ref: Guideline on the investigation, management and prevention of venous thrombosis in children. *Brit Journal of Haematol*;2011(154):196–20.

On C&V Web, the full guideline includes appendices on:

A. Enoxaparin preparation and home use
B. Paediatric Warfarin treatment chart
C. Surgical thromboprophylaxis risk assessment

Loading with warfarin

Refer to the all-Wales anticoagulation prescription chart.

- Review the indication for warfarin – confirm desired range with consultant
- Obtain a baseline INR – a full coagulation screen is needed if the patient is on heparin*
- Load as below and obtain daily INRs for the first 5 days – patients should be dosed under the supervision of paediatric cardiologist/paediatric haematologist
- Heparin should continue until INR is in target range for 2 consecutive days

Remember to check baseline PT/INR. If PT is ≥1.3, the patient will need a lower loading dose.

The recommended loading dose is 0.1 – 0.2 mg/kg (lower dose for Fontan patients or those with impaired liver function; higher dose for other patients). In practice the following guide may be applied:

Age group	Loading dose	Comment
Infants (6-12 months)	1 – 2 mg	Check INR next day
Young children (1-5 years)	3 – 5 mg	Check INR next day
Older children (5-12 years)	5 – 8 mg	Check INR next day
Teenagers	8 mg	Check INR next day
Adults	Use the All Wales adult warfarin chart	

*In patients on unfractionated heparin who are being loaded on warfarin, it is essential to send a full coagulation screen. A high KCCT/APPT will lead to an erroneously high INR level. If only the INR is checked, the true reading off heparin may be much lower.

6.1.7 INR Sampling

It is the responsibility of the person taking the blood sample for INR estimation to find out the result and prescribe the appropriate dose of anticoagulant (see INR protocol below).

A portable INR machine is kept on the ward and is available for the use of paediatric cardiac patients. Only staff who have been trained appropriately are authorised to use it.

If an INR reading is unexpectedly deranged, consider the cause (intercurrent illness, drug interaction, alcohol ingestion).

6.1.8 INR Protocol

Use this protocol when prescribing warfarin. If unsure, check with consultant. (see Warfarin Dosage Table in section 6.1.9). From 2013, “new entrants” <8 years and current patients whose INR control is deemed “unstable” will be prescribed commercial warfarin liquid. Remember that the target INR for prosthetic valves is 3.0 (unless specified otherwise) and the target INR for most other indications is 2.5.

THE INR DOSING TABLES ARE SUITABLE FOR USE IN PATIENTS WITH A RELATIVELY STABLE INR – PATIENTS WITH SHORT OR SUSTAINED PERIODS OF POOR CONTROL NEED TO BE CONSIDERED BY SENIOR STAFF ON AN INDIVIDUAL BASIS.

PATIENTS WITH A “UNIQUE” TARGET INR DOSE WILL HAVE AN INDIVIDUALISED DOSAGE TABLE APPROVED BY THEIR CONSULTANT.

Patients will be prescribed warfarin in tablet or liquid form. See tables below to guide dose prescription.

6.1.9 Warfarin tablet dosage table (doses shown in mg)

10	5
9.5 / 10 / 10	4.5 / 5 / 5
9.5 / 10	4.5 / 5
9.5 / 9.5 / 10	4.5 / 4.5 / 5
9.5	4.5
9 / 9.5 / 9.5	4 / 4.5 / 4.5
9 / 9.5	4 / 4.5
9 / 9 / 9.5	4 / 4 / 4.5
9	4
8.5 / 9 / 9	3.5 / 4 / 4
8.5 / 9	3.5 / 4
8.5 / 8.5 / 9	3.5 / 3.5 / 4
8.5	3.5
8 / 8.5 / 8.5	3 / 3.5 / 3.5
8 / 8.5	3 / 3.5
8 / 8 / 8.5	3 / 3 / 3.5
8	3
7.5 / 8 / 8	2.5 / 3 / 3
7.5 / 8	2.5 / 3
7.5 / 7.5 / 8	2.5 / 2.5 / 3
7.5	2.5
7 / 7.5 / 7.5	2 / 2.5 / 2.5
7 / 7.5	2 / 2.5
7 / 7 / 7.5	2 / 2 / 2.5
7	2
6.5 / 7 / 7	1.5 / 2 / 2
6.5 / 7	1.5 / 2
6.5 / 6.5 / 7	1.5 / 1.5 / 2
6.5	1.5
6 / 6.5 / 6.5	1 / 1.5 / 1.5
6 / 6.5	1 / 1.5
6 / 6 / 6.5	1 / 1 / 1.5
6	1
5.5 / 6 / 6	0.5 / 1 / 1
5.5 / 6	0.5 / 1
5.5 / 5.5 / 6	0.5
5.5	0 / 0.5 / 0.5
5 / 5.5 / 5.5	0 / 0.5
5 / 5.5	0 / 0 / 0.5
5 / 5 / 5.5	

NB 5 / 5 / 5.5 means give 5 mg Day 1, 5 mg Day 2 and 5.5 mg Day 3; then repeat the cycle

6.1.10 Warfarin prescription - patients with prosthetic valve (tablet or liquid) (assuming target INR is 3.0)

INR	Action/Comment (Tablet)	Action/Comment (Liquid)
2.0 or below with prosthetic valve	(1) Inform on call consultant. (2) Give early (i.e. morning) dose of warfarin (3) Admit patient for Clexane (4) Urgent echocardiogram to review status of the valve. (5) Check INR 3.30 pm to determine evening dose of warfarin and the need for further Clexane doses (6) Arrange to repeat the INR the next morning. Anticipate needing to go up 3 increments on warfarin dosage table (6.1.9)	See column left. Once actions 1-5 have been taken, arrange to repeat the INR the next morning. Anticipate needing to increase the warfarin dose by 15%.
2.1 to 2.4	(1) Inform on call consultant. (2) Consider the need hospital review for Clexane and to check valve status (3) Consider need for extra morning dose of warfarin; if given, recheck INR again in the evening; (4) Refer to table 6.1.9. Move warfarin dose up the table by 2-3 increments depending on the trend.	See column left Once points 1-3 have been actioned, anticipate needing to increase the warfarin dose by 10-15% depending on the trend.
INR 2.5 to 2.8	Refer to table 6.1.9. Keep warfarin dose the same or move warfarin dose up the table by 1 increment depending on the trend.	Either maintain current warfarin dose or increase by 5% depending on the trend.
INR 2.9 to 3.1	No change in dose.	No change in dose.
INR 3.2 to 3.5	Refer to table 6.1.9. Keep warfarin dose the same or move warfarin dose down the table by 1 decrement depending on the trend.	Either maintain current warfarin dose or reduce by 5% depending on the trend.
INR 3.6 to 3.9	Refer to table 6.1.9. Move warfarin dose down the table by 1-2 decrements depending on the trend.	Reduce the dose of warfarin by 5-10% depending on the trend.
INR 4.0 to 5.0	Refer to table 6.1.9. Move warfarin dose down the table by 3 decrements. Recheck in 1-2 days.	Reduce the warfarin dose by 15%. Recheck in 1-2 days.
INR 5.1 to 5.9	Inform consultant. Reduce dose by 50% or more, or withhold dose, recheck next morning	
INR 6.0 or above	Refer to advice below regarding high INR	

6.1.11 Warfarin prescription – most other patients (e.g. Fontan, DCM, thrombosis, assuming target INR is 2.5)

INR	Action/Comment (Tablet)	Action/Comment (Liquid)
<1.5	See table 6.1.9. Move dose up the table by 3 increments.	Increase warfarin dose by 15%.
1.5 to 1.9	See table 6.1.9. Move dose up the table by 1-2 increments depending on the trend.	Increase warfarin dose by 5-10%, depending on the trend.
2.0 to 3.0	No change	No change
3.1 to 4.0	See table 6.1.9. Move the dose down the table by 1-2 decrements depending on the trend.	Reduce warfarin dose by 5-10%, depending on the trend.
4.1 to 5.9	Stop Warfarin, restart on a reduced dose (3 decrements down the table) when INR is <3.1.	Stop warfarin, restart on dose 15% lower once the INR is <3.1.
>5.9	Refer to advice below regarding high INR	

Recheck INR (see also Table 6.1.9 for patients on tablet warfarin):

- Stable (and no worrying trend up or down) 4-6 weeks
- Dose changed 1 place (or 5%) up or down 1-2 weeks (depending on trend)
- Dose changed >1 place up or down (or 10-15%) 1 week (or less, depending on trend)
- Dose stopped, halved or undergoing loading Next day

6.1.12 Management of low INR readings – general principles

In patients on warfarin for reasons other than prosthetic valve, unless there is an active problem with thrombosis (i.e. there is a known clot in the circulation), it is usually not necessary to use heparin or Clexane. The dose of warfarin should be increased as per protocol and the INR rechecked in 5-7 days.

In patients with a prosthetic valve:

- Inform the on-call consultant
- Advise give early (i.e. morning) dose of warfarin
- Consider need for urgent echocardiogram
- Consider the need for subcutaneous Clexane (1mg/kg)
- Consider need for hospital assessment of prosthetic valve (± admission)
- Arrange for INR to be rechecked 3-3.30 pm
    - Consider dose of warfarin to be given in the evening
    - Consider whether second dose of Clexane should be given (12 hours after the first)
- Arrange for INR to be rechecked the following morning and prescribe appropriate dose of warfarin

6.1.13 Management of high INR readings

ADC 2011;96:164-7 provides evidence that if the INR reading is >5 and <8 and there is no active bleeding, then omitting the warfarin until the INR is <5 is safe and that no vitamin K is needed. In general, the following principles should be followed:

1) Patients with **prosthetic heart valves** should not be given intravenous vitamin K (reconsider if major bleeding).

2) For **all other patients** the following recommendations are adapted from the British Society for Haematology. If you are unsure, consult a haematologist (or paediatric cardiologist or surgeon if the patient has a prosthetic heart valve).

INR in therapeutic range – patient bleeding

- Investigate source of bleeding. Consider risk/benefit of stopping warfarin.

INR < 6.0 but > 0.7 above target INR – no bleeding

- Reduce the dose following the advice on maintenance dosing (see relevant table above)

INR > 6 – no bleeding or minor bleeding from mucosa (nose, oropharynx, urinary tract, rectum, anus)

- Stop warfarin
- Restart when INR < 5.0
- Assess patient for their risk of bleeding: recent surgery/trauma, extensive bruising, minor mucosal bleeding
    - If at high risk of bleeding, give vitamin K 2 mg orally
    - Use Konakion MM paediatric® (phytomenadione 2 mg in 0.2 mL)
    - Draw up using oral dispenser provided and then drop onto the tongue.
- Recheck INR after 24 hours, and consider need for repeat dose of Vitamin K if INR is still too high.

Major bleeding: Life or limb threatening bleeding, including intracranial haemorrhage

- Stop warfarin
- Give 250-300 microgram/kg (max 10mg) vitamin K IV (Phytomenadione 10mg/mL – Konakion MM®.) Give as an IV bolus over 3-5 minutes undiluted or diluted with 10-20mL of glucose 5% to aid slow administration.
- Give prothrombin complex concentrate* (PCC - Factor II, VII, IX and X concentrate) obtained from blood bank (not pharmacy) – dose to be advised by haematologist. Dissolve in water for injection as per manufacturer's guidance, using an aseptic technique and the provided transfer device. Administer over 10 minutes. See local protocol for further details on administration (should be provided from blood bank with the prothrombin complex concentrate).
- Repeat INR within 1 hour of giving of PCC – consider further dose if INR remains >1.5 and patient still bleeding (discuss with haematologist).
- Consider risk-benefit of recommencing warfarin.

*PCC dose: INR 2-3.9 [25 U/kg]; INR 4-5.9 [35 U/kg]; INR >6 [50 U/kg]

Contributors: Philip Connor, Claire Logan, Orhan Uzun, Elizabeth Webb, Dirk Wilson

6.1.14 Management of bleeding in patients on DOAC therapy

A small number of paediatric patients are on the direct oral anticoagulant therapies (e.g. Apixaban, Rivaroxaban). This form of treatment is not given to patients with prosthetic valves and brief cessation of therapy, if needed, is unlikely to result in a significant thrombotic complication.

If a patient on this form of treatment has a bleeding complication the following advice applies:

- Minor bleeding – stop treatment and investigate source of bleeding
- Major bleeding – discuss with the on-call Haematologist. Discuss use of prothrombin complex concentrate* (PCC - Factor II, VII, IX and X concentrate) at a dose of 25-50 U/kg. PCC is obtained from blood bank (not pharmacy) – dose to be advised by

haematologist. Dissolve in water for injection as per manufacturer's guidance, using an aseptic technique and the provided transfer device. Administer over 10 minutes.

- Discuss with Haematology about possible use of Andexanet alpha. There are very little data in children, but this may be of some benefit if bleeding continues to be an issue.
- Please note - Vitamin K will not have any effect on DOAC reversal or levels and would therefore not be recommended.

See:
https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6913453/#:~:text=Bleeding%20is%20the%20main%20complication%20of%20anticoagulant%20therapy&text=2%2D6-,Although%20direct%20oral%20anticoagulants%20(DOACs)%20reduce%20the%20risk%20of%20major,patients%20who%20have%20major%20bleeds.

6.1.15 Cessation of warfarin for surgical or invasive procedure

Generally speaking it is safe to undertake a "minor" procedure (cardiac catheterisation, dental extraction, minor non-cardiac surgery) with an INR of 2.2-2.4 or less. Where the type of surgery is associated with a high risk of bleeding (e.g. spinal surgery, liver surgery or biopsy, renal biopsy, cranial surgery) an INR of <1.6 is desirable.
Patients with prosthetic heart valves* need careful peri-operative management. The following approach is recommended, although if the risk of operative bleeding is low it may be safe to proceed with the operation on a slightly reduced dose of warfarin:

- Check INR 3 days before planned procedure (the parents can do this if they have a home INR monitor)
- If the INR was in therapeutic range, give the normal dose 3 days pre-op (if not, discuss with consultant)
- Discontinue warfarin therapy 2 days before procedure and admit to hospital
- Repeat INR daily
- If INR falls below 2, start iv heparin therapy (including loading bolus) with the aim of maintaining APTT/KCCT ratio 1.5-2.0
- Discontinue heparin therapy 6 hours before surgery
- Recommence heparin (with reloading bolus if surgery was lengthy) once bleeding is controlled; aim for APTT/KCCT ratio 1.5-2.0
- Recommence warfarin (consider gentle loading dose) and maintain heparin until INR is therapeutic for 24-48 hours
- Remember – a full lab coagulation screen is needed for patients on heparin and Warfarin
- Restart warfarin the next day. Reloading may be required.

NB: Clexane may be used to replace continuous iv heparin in some circumstances – stop warfarin and arrange for admission as above. Once INR is <2.0, give Clexane 1 mg/kg/dose every 12 hours. Give final dose of heparin 24 hours before planned surgery. Restart Clexane once the risk of serious bleeding has passed. Restart warfarin the next day. Reloading may be required.

*Please note that patients who are anticoagulated for dilated cardiomyopathy or following a Fontan procedure generally do not require the above steps in preparation for invasive procedures – warfarin therapy can be stopped ~2 days prior to the procedure. It is advised to check the INR prior to the procedure ensuring the reading is <2.2-2.4, then normal therapy can be resumed afterwards.

6.1.16 Factors that influence the efficacy of warfarin

Patient factors:

- Enhanced anticoagulation effect with

    - Weight loss
    - Intercurrent illness
    - Liver disease
    - Heart failure
    - Renal failure
    - Excess alcohol ingestion

- Reduced anticoagulant effect
    - Weight gain
    - D+V
    - Asian or Afro-Caribbean background

Drug interactions with warfarin:

- Reduced protein binding/inhibition of other coagulation pathways
    - Aspirin or other anti-platelet drugs
    - Chlorpromazine

- Inhibition of warfarin metabolism
    - Erythromycin
    - Sodium valproate
    - Cimetidine
    - Cranberry juice

- Enhanced metabolism of warfarin
    - Phenytoin
    - Carbamazepine
    - Phenobarbitone

- Reduced synthesis of coagulation factors II, VII, IX and X
    - Phenytoin
    - Salicylates

- Reduced absorption of vitamin K
    - Broad-spectrum antibiotics
    - Laxatives

Enhanced risk of bleeding is seen with aspirin and other NSAIDs, corticosteroids, thrombolytics. Consider prescribing ranitidine to reduce the risk of GI bleeding.

Monitor INR more closely if new medications are prescribed, or if the patient is acutely unwell.

(Source: ABC of antithrombotic therapy. BMJ 2002;325:762-4 + advice from Dr P Connor).
Contributors: Philip Connor, Clara Danielson, Claire Logan, Nav Masani, Orhan Uzun, Dirk Wilson

6.2 Asplenia and Immunodeficiency

It is not uncommon for cardiac patients to have specific immune problems. If there is any doubt in dealing with immunosupressed patients, consult with the Public Health (Virology) and Paediatric Infectious Disesase / Immunology Departments.

6.2.1 Asplenia

This occurs in cardiac children with right atrial isomerism (known as “asplenia syndrome in the USA). Children with left atrial isomerism (“polysplenia syndrome”) may also have functional hyposplenism and this needs to be assessed carefully.

Children with absent or dysfunctional spleen tissue are at risk of overwhelming infection particularly with encapsulated organisms such as *Pneumococcus*, *Meningococcus* and *Haemophilus influenza*. Other high risk infections in this group include malaria, *E. coli*, *Babesiosis* (tick borne disease) and infections transmitted by animal bites.

A departmental audit recommended the following for children with suspected isomerism of the atrial appendages:

- Define atrial morphology and cardiac abnormalities
- Define anatomical splenic status
- Define abdominal viscera morphology
- Define bronchial morphology
- Refer to immunologist (regardless of anatomical splenic status)
- Begin routine vaccinations
- Initiate prophylactic antibiotics
- Determine functional splenic status (at 4 months)

Asplenic children under 5 years have a particularly high infection risk of overwhelming sepsis (>10%). The following antibiotic prophylaxis is recommended:

No penicillin allergy:

Age	Antibiotic
1 mo – 4 years	Amoxicillin 125mg twice daily
5 – 11 years	Amoxicillin 250mg twice daily
12 – 18 years	Amoxicillin 500mg twice daily

Alternative if penicillin allergic:

Age	Antibiotic
1 mo – 23 mo	Erythromycin 125mg twice daily
2 – 7 years	Erythromycin 250mg twice daily
8 – 17 years	Erythromycin 500mg twice daily

Advice for family:

- Parents should be made aware of the excess infection risk and the need for immediate treatment of suspected infection.
- They should have a reserve supply of antibiotics at therapeutic doses to take on holiday.
- They should be given the Asplenia Warning Card to carry.
- The excess risk from malaria should preclude unnecessary travel to endemic areas.
- Antibiotics should be given after animal bites (Augmentin) and despite prophylactic antibiotics a serious suspected infection should be given immediate treatment with cefotaxime or ceftriaxone.
- Isomerisms can be familial therefore consider screening other family members.

Childhood immunisations – speak to the paediatric infectious diseases team and refer to the “Green Book” – see:

https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/857279/Greenbook_chapter_7_Immunsing_immunosupressed.pdf

References:

1 – Davies JM, Barnes R, Milligan D. Update of guidelines for the prevention and treatment of infection in patients with an absent or dysfunctional spleen. *Clin Med.* 2002 Sep-Oct; **2(5):** 440-3.
2 – Finn A, Booy R, Moxon R, Sharland M, Heath P. Should the new pneumococcal vaccine be used in high-risk children? *Arch Dis Child.* 2002 Jul; 87(1): 18-21. Review.
3 – https://www.gov.uk/government/publications/immunisation-of-individuals-with-underlying-medical-conditions-the-green-book-chapter-7

Contributors: Siske Struik, Orhan Uzun, Dirk Wilson

6.2.2 DiGeorge Syndrome (22q11.2 deletion syndrome)

DiGeorge syndrome is caused by a deletion on the long arm of chromosome 22 (in full: 22q11.2 deletion syndrome) and is broadly associated with midline defects. It has a broad spectrum of clinical features, not all of which may be present in any individual patient, and severity varies widely also within affected families, i.e. variable penetrance and phenotype. Not all children with 22q11.2 deletion syndrome have cardiac defects.

Whilst not well known in the public domain, it is actually common and with increased genetic testing in the context of developmental delay pick up is increasing. Current estimated prevalence is 1 in 2000-4000 births. For comparison, Down syndrome prevalence in the UK is currently estimated at 1:4000.

Depending on how they are affected children will present early, or much later. Indeed it is likely a significant proportion of young people and adults remain undiagnosed.

Key early presenting features include:

- Characteristic facial appearance (subtle midline features)
- Congenital abnormality of the heart or great vessels (particularly conotruncal abnormalities, these should trigger cardiologists to send an array to genetics promptly – see section 5.1.2)
- Hypoparathyroidism (presenting as hypocalcaemia with inappropriately low PTH)
- Velopharyngeal dysfunction (with variable swallowing problems and speech and language development difficulties)
- Absent thymus gland with low T cells and reduced immune function (typically frequent respiratory infections with gradual improvement with age)
- Renal/urological abnormalities and many other congenital abnormalities are rarer but can be associated
- A variable degree of developmental delay, social communication and emotional difficulties is also associated, and only will become apparent over time.

Inheritance/genetics

Autosomal dominant. Approximately 90% are new mutations, with 10% inherited (parents often undiagnosed). Therefore once identified in a child, prompt referral to Clinical Genetics for family assessment is recommended, both to identify health care needs of other affected family members and to counsel parents for recurrence risk in future pregnancies.
It is increasingly recognised that children with a duplication, as opposed to deletion, of 22q11 have similar phenotypes and investigation and management upon this diagnosis therefore follows the same pathway initially.

Infections/Immunology
Increased susceptibility to infection, and increased morbidity with normal childhood viral infections, is common, and often not purely for immunological reasons, but in combination with cardiac compromise, post-surgery recovery, reduced efficiency of chest clearance (unwell, and generally slightly lower muscle tone), and swallowing difficulties with associated aspiration risk.
However, all new diagnoses of 22q microdeletion should have a Paediatric Immunology work up (blood tests) and clinical review. Many will require long term outpatient Paediatric Immunology follow up.

All Wales Paediatric 22q MDT clinic
This clinic is held in UHW 3 times a year. This clinic sees all new diagnoses at least once, usually within 4-6 months of the diagnosis, to review the phenotype comprehensively, advise, refer and signpost families to the required services and peer support networks. This clinic is not for acute new problems, which should be managed and referred by relevant specialist directly.
The current clinic MDT consists of Paediatric Immunology, Cleft surgeon, specialist 22q speech and language development therapist and psychologist (note this is separate from swallowing problems, which are dealt with by local SLT teams), and genetics counsellor.
Referrals to the MDT clinic can be made in writing to Paediatric Immunology who will often see families first in their clinic and then book into MDT clinic at an appropriate time frame for the family.

When children are diagnosed with 22q11 deletion syndrome via the cardiac route:

1. Please look at the following link 22q11 Paediatric Service - Cardiff and Vale University Health Board (nhs.wales)
    - This includes a prompt list for baseline blood tests and renal ultrasound, which you will need to organise and action if abnormal (e.g. endocrine, cleft service, or renal referrals)
2. All new diagnoses have an outpatient referral made to Paediatric Immunology and this link explains how and what to include in the letter.
3. Unwell inpatients with a cardiac diagnosis, problems with infections and a new diagnosis of 22q11 microdeletion syndrome should be reviewed by Paediatric Immunology as inpatients (consultant to consultant request). Stable in/outpatients can be referred by letter, provided all investigations have been initiated.
4. Pending Paediatric Immunology review, particularly in unstable infants:
    - Hold off live vaccines (rotavirus, MMR, BCG)
    - If transfusion required, give irradiated CMV negative blood (and inform blood bank so they can put a note on their system)
    - Manage chickenpox contacts as per Green Book instructions (assume immunocompromised)
    - Ensure household members up to date with all vaccinations including Flu (intranasal live Flu for siblings OK, unless patient very unwell, in which case advise IM inactivated Flu)
    - Give Palivizumab (this is safe) as per normal cardiac guidelines
    - Ensure review by Paediatric Immunology prior to discharge so that Co-trimoxazole prophylaxis can be considered and prescribed.
5. Please give a brief explanation to the family that 22q11 microdeletion syndrome has been identified. The following link is to the (excellent) patient organisation. It has very useful information for professionals and families alike:
   Max Appeal (www.maxappeal.org)
   Please take a good look yourself at this website and tailor your initial information to the immediate needs of the family, which are likely focussed on coping with the cardiac issue. It is important to discourage googling and rely on the given link which has lots of reliable information. Also stress that whilst this diagnosis can be associated with

many different problems, individuals vary widely. The urgent issues (cardiac, feeding, infections, calcium, renal) need addressing first. In time, more detailed reviews will happen and it will gradually become clearer how their child is affected.

6.3 Bradycardia and Normal Heart Rate in the Newborn Period

The normal heart rate increases from the first day of life. It reaches a peak between the first and the second month and then declines returning to the values recorded at birth by the sixth month. During the following 6 months, it remains stable and then slowly declines after 1 year due to maturation of vagal innervation of the sinus node.

Clinically significant gender differences in heart rate are not seen in the neonatal period. Resting and sleeping heart rates in newborns and infants are lower compared to when they are alert and heart rate increases significantly when they are crying.

Mean heart rate in the first year of life range increases from 123 beats per minute (bpm) at birth, reaching a maximum of 150 bpm at about 1 month of age, before decreasing to 113 bpm by age 2 years. Heart rates between the 2nd and 98th percentile in the first year of life are shown in below.

Age	Heart rate 2^{nd} to 98^{th} percentile in bpm (mean)
0-1 days	93-154 (123)
1-3 days	91-159 (123)
3-7 days	90-166 (129)
7-30 days	107-182 (140)
1-3 months	121-179 (150)

Some causes of neonatal bradycardia:

1) Sinus bradycardia:

- Hypoxia
- Acidosis
- Infection / sepsis
- Electrolyte abnormalities
- Neonatal hypothyroidism
- Increased intracranial pressure
- Hypervagal states- e.g. high position of NG tube, Gastro oesophageal reflux disease
- Obstructive jaundice

2) Sinus node dysfunction - consequence of abnormal development or direct injury to sinus node

- Central line tip in right atrium
- Congenital heart disease (atrial isomerism, ASD, AVSD, single ventricle, CCTGA)
- Post cardiac intervention (e.g. cardiac catheterisation, surgery)

3) Conduction abnormalities or channelopathy

- Kearne-Sayre Syndrome
- Long QT syndrome

4) Heart block

- Congenital- maternal connective tissue disorders

- Acquired- post-surgery, myocarditis, rheumatic heart disease, congenital syphilis, diphtheria, Lyme disease.

Recommendations:
Persistent neonatal bradycardia in an awake baby of less than 90 beats per minute (2nd percentile) in the newborn period should prompt assessment and investigation to rule out common cardiac and non-cardiac causes of bradycardia. A careful assessment of the baby needs to be undertaken by the neonatal team before referral to cardiology. The assessment should include:

- History
    - Maternal drug history
    - Maternal connective tissue disorders
    - Birth details – difficult labour resulting in hypoxia
- Examination
- Blood tests
    - Glucose and capillary blood gas
    - Serum electrolytes, renal function, calcium and magnesium
    - Follow NNU thyroid function test guideline (in addition to Guthrie card)
    - SS-A (anti-Ro antibodies) and SSB (anti-La antibodies)
- Baseline ECG with calculation of QTc and PR interval (by the ECG computer package – confirm with hand calculation if abnormal)

If the ECG is abnormal, or any abnormality is found in the cardiovascular examination or if the baby becomes unwell, discussion with on-call paediatric cardiology team should be undertaken for urgent assessment including echocardiography.

The baby should be observed by the neonatal/midwifery transitional care team for 2 days pending results of any investigations. At least 4-hourly monitoring is recommended. A repeat ECG should be performed on day 3. If the heart rate has normalised and all the investigations are normal then the baby can be discharged home. If there is medical or parental concern, consider the need for neonatal clinic follow-up.

If the heart rate remains <2nd percentile in an awake baby, all the investigations are normal and the baby remains well then a referral to Paediatric Cardiology should be made. An urgent 24 hour Holter should be considered.

References:

1. Fleming S, Thompson M, Richard S et al. Normal ranges of heart rate and respiratory rate in children from birth to 18 years of age: a systemic review of observational studies. *Lancet* 2011; 377 (9770), 1011-18.
2. Schwartz PJ, Garson Jr A, Paul T et al. Guidelines for the interpretation of the neonatal electrocardiogram, A Task Force of the European Society of Cardiology. *Eur Heart J* 2002; 23: 1329–44.
3. Michelle S. Miller, Kevin M. Shannon, Glenn T. Wetzel. Neonatal Bradycardia. Progress in Pediatric Cardiology, Elsevier, 11 Ž2000. 19.24

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6.4 Cardiac Failure

Definition – Heart failure is a complex clinical syndrome which results from any structural or functional impairment of ventricular **filling**, or the **ejection** of blood. There is a mismatch between the metabolic demand of the body vs ability of the cardiovascular system to meet that demand.

This may arise from disorders of

- Pericardium
- Myocardium (see section 6.5 Cardiomyopathy)
- Endocardium
- Valves
- Great vessels

Aetiology:

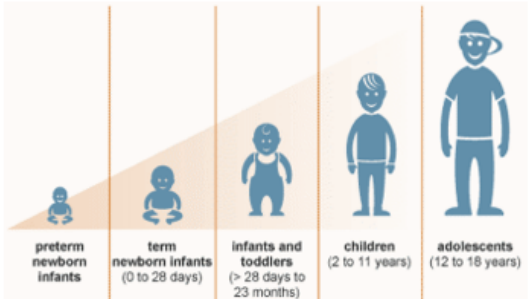
Newborns and Infants	CHD	L→R shunt
		L heart obstruction
		Single ventricle, valve leak
	Heart muscle disease	Myocarditis
		DCM, HCM
	Coronary abnormality	ALCAPA
		KD
	Sustained arrhythmia	AT, AVRT, VT
	Inborn errors of metabolism	Storage disorders, Barth, Mitochondrial cytopathy
Age 2-5 years	Heart muscle disease	Myocarditis
		DCM
	CHD	Single ventricle, valve leak
	Sustained arrhythmia	AT, AVRT, VT
	Coronary abnormality	KD
Age >5 years	Heart muscle disease	Myocarditis
		DCM, HCM, ARVC
	Sustained arrhythmia	AT, AVRT, VT
	Inflammatory disease	Rheumatic fever, SLE
	Endocrine	Thyroid, Phaeo
	Haematological	Anaemia, Sickle, Thallass
	Toxins	Anthracylines, Radiation
	Renal	Hypertension

Pathophysiology:

- CVS insufficiency → neurohormonal activation
  - Sympathetic nervous system
  - Renin–angiotensin–aldosterone system
- Neuro-hormonal activation → coordinated responses that work to restore cardiovascular homeostasis in the short term
- Sustained neuro-hormonal activation drives the progression of deleterious effects on the circulation and the myocardium

Presenting features:

Tachypnoea
Feeding difficulty (reflux, vomiting, feeding refusal)
Growth failure
Sweating
Pallor



Fatigue
Effort intolerance
Dyspnoea
Orthopnoea
Abdominal pain
Nausea
Vomiting

Clinical manifestations include

- Breathlessness
- Fatigue
- Fluid retention
    - Pulmonary
    - Splanchnic
    - Peripheral

Symptomatic severity:

Functional class	Symptoms noted on history
I	Asymptomatic
II	Infants: mild breathlessness or sweating with feeding – no growth failure
	Older children: breathless on moderate exertion
III	Infants: marked breathlessness or sweating with feeding – growth failure
	Older children: breathless on mild or minimal exertion
IV	Breathlessness, sweatiness at rest

Infants: 0-1 year
Older children >1 yr

Management
<u>History</u>

- Timing of symptoms
- Severity
- FHx (3 generation)
- Seek clues as to aetiology

<u>Examination</u>

- Baseline observations
- Effect on feeding and growth
- Peripheral circulation
- Impact on respiratory system
- Evidence of fluid retention
- Seek clues as to aetiology

Parameter	Adequate CO	Inadequate CO
Heart rate	Normal	elevated
Blood pressure	Normal	normal or low
Arterial waveform	large area under curve; late dicrotic notch	small area under curve; early dicrotic notch
Pulses	peripheral pulses easy to feel	peripheral pulses weak central pulses palpable
Peripheral perfusion	capillary refill < 2sec	capillary refill >2sec
Core-toe temperature gap	< 2^{o}C	> 2^{o}C
Urine output	>1 ml/kg/hr	<1 ml/kg/hr
Base deficit	<2 mmol/L	>2mmol/L
Mental status	co-operative	agitated, disorientated

Investigation

- ECG
- Chest x-ray
- Echocardiogram
- Oximetry
- Capillary gas (if unwell)
- Holter
- Urine testing (rule out metabolic disease, test urine VMA)
- Blood tests
    - FBC + film
    - U&E/LFT
    - TFT
    - ASOT
    - Serum virology
    - CRP/ESR
    - Troponin
    - Autoantibodies
    - pro-NT-BNP

Treatment

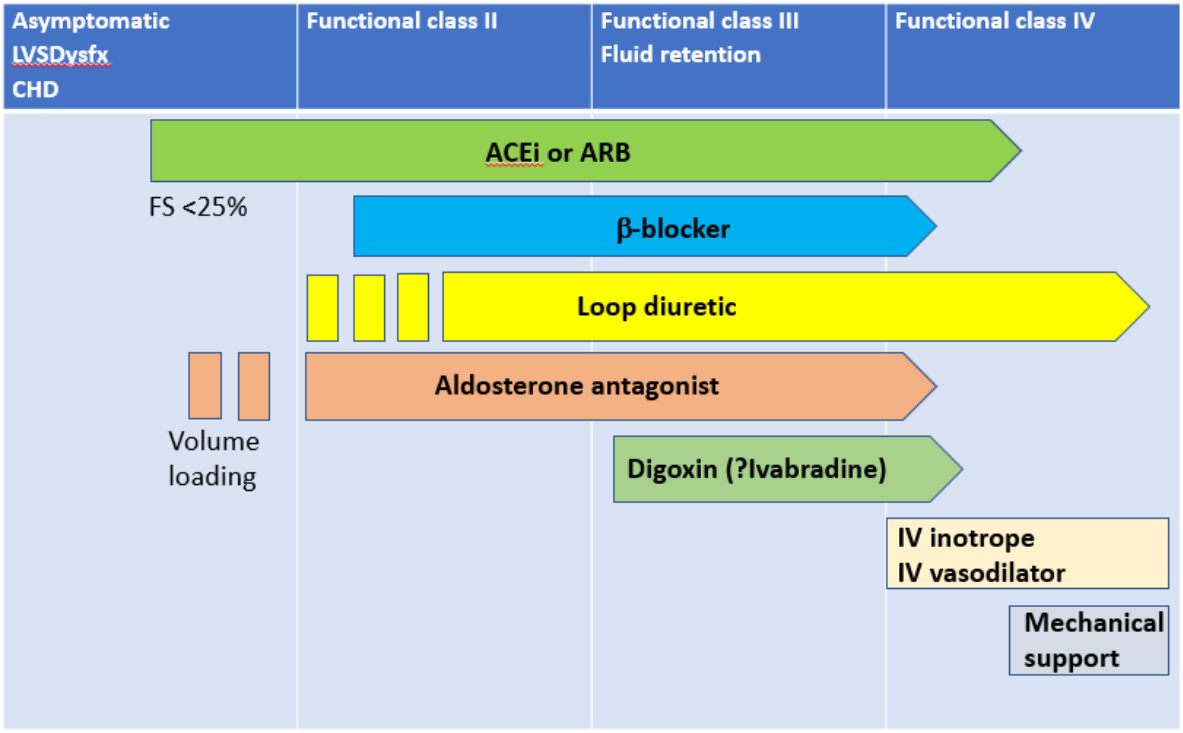
- Oxygen to maintain saturation (beware that excessive use can make heart failure worse in left to right shunts)
- Consider sedation in the distressed child
- Treat fluid overload – fluid restriction and diuretics
- Prevent further salt and water retention – diuretics, angiotensin converting enzyme (ACE) inhibitors, and angiotensin receptor blockers
- Tachyphylaxis – β-Blockers and digoxin (maintain digoxin level at 0.5-1.0ng/L)
- Increase myocardial contractility – Inotropes or inodilators (PDE inhibitors preferably) and digoxin

Aims of Treatment

- Acute HF management – aims
    - Improve haemodynamics
    - Prevent progression
- Chronic HF management – aims
    - Maintain stability
    - Prevent progression

- Provide sufficient symptomatic relief to allow optimal growth and development
- Consider interventions that may reduce mortality

Heart failure in children – medical therapy



Catecholamines table

Drug	β1	β2	α	Dopaminergic
Dopamine	++		++	+++
Dobutamine	+++	+	+	
Isoprenaline	++++	++++		
Adrenaline	++	++	+++	
Noradrenaline	++		++++	

Phosphodiesterase inhibitors:
Milrinone amrinone, and enoximone are phosphodiesterase inhibitors that increase intracellular cyclic AMP. They increase peripheral vasodilation and improve ventricular relaxation allowing increased ventricular filling. Milrinone is the agent most commonly used in paediatric practice. It is given via continuous IV infusion – refer to BNFc or local PICU or NICU guideline for prescription advice.

Catecholamines, PDE inhibitors and cardiac glycosides improve myocardial contractility at different levels of intracellular calcium transport. Their receptor actions are however not specific and they thus have multiple effects.

After-load may become a critical factor determining myocardial function because the autonomic response to a poor cardiac output is an increase in systemic vascular resistance, and hence after-load, further compounding the problem. Reduction in after-load thus reduces

myocardial work and oxygen requirement and improves cardiac output**.** Suitable drugs include glyceryl trinitrate, sodium nitroprusside, prostacyclin, phenoxybenzamine and ACE inhibitors.

Too slow a rate will reduce the cardiac output. The rate can be increased with the use of isoprenaline or pacing. Ideally if pacing is used, sequential A-V pacing (atria then ventricles) should be selected.

Sometimes the cardiac rate may be too high (> 200 bpm) to allow adequate ventricular filling. Digoxin or β-blockers may be used to reduce the rate by slowing AV node conduction.

Advanced therapies:

- Entresto (Sacubitril with Valsartan) – not licensed in children, but research trials are underway. Licensed in the USA – see: https://www.accessdata.fda.gov/drugsatfda_docs/label/2019/207620s013lbl.pdf
- Biventricular pacing (CRT)
    - Benefit shown in adults with
        - LBBB and QRSD >150 msec
        - LVEF 35% or under
        - Symptomatic
- Indications in children
    - Symptomatic DCM with broad QRS and severe LV dysfunction despite medical therapy
    - Pacing-induced cardiomyopathy
- Leads can be placed epicardially in smaller children
- Implantable Cardioverter-Defibrillators
    - Lack of data and clinical experience – no published recommendations
    - In adults with DCM, primary prevention ICD implant is recommended with
        - NYHA class II-III
        - Ejection fraction (EF) ≤35%
- Can be combined with biventricular pacing (CRT-D)

End-stage heart failure options:

- Advanced medical heart failure management combined with palliative care
    - IV inotropes and vasodilators – value as a bridge to transplantation
- Mechanical circulatory support
    - ECMO
    - Ventricular assist device (VAD)
- Heart transplantation – see section 6.29

Further reading/References

CVPhysiology.com
Curr Cardiol Rev 2016;12(2):99-103
https://www-nature-com.abc.cardiff.ac.uk/articles/nrcardio.2016.163
Paediatrics in Review 2019;40:60-70
Can J Cardiol 2013;29:1535-52
https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6069285/

Contributor: Dirk Wilson

6.5 Cardiomyopathy

In most children with dilated cardiomyopathy an underlying cause is not identified. However there are several rare metabolic disorders that may be associated with cardiomyopathy, and, whilst certain conditions may be suggested by the clinical presentation, not uncommonly

metabolic conditions may have no extra-cardiac features at all. It is therefore very important that a carefully structured investigation protocol is followed to prevent these being overlooked.

Aetiology

- Infective
- Metabolic & cellular
- Immunological
- Toxins (e.g. anthracylines, alcohol)
- Left heart obstruction (e.g. AS, CoA)
- Hypertension
- Coronary abnormalities (anomalous LCA)
- Chronic left-to-right Shunt
- Intractable arrhythmia
- Familial (~20%)
- Peripartum

History

- Parental consanguinity
- Antenatal history - HELLP Syndrome, Fatty Liver of Pregnancy or Severe PET, consider fatty acid oxidation defect
- Neonatal death or SIDS in a sibling
- Symptoms of preceding viral infection
- Liver disease or multiple transfusions
- Myopathy, ataxia or autoimmune disease
- Abnormal nutrition history (e.g. long term parenteral nutrition)
- Delayed development
- Past medical history, malignancy, chemotherapy (cumulative dose of anthracyclines)
- Myocardial ischaemia may have an unusual presentation in children who are unable to localise pain
- Draw a detailed family tree – pay close attention to relatives with heart muscle problems, heart rhythm disturbance and sudden cardiac death, and a history of recurrent miscarriage, stillbirth or early neonatal death.

Examination

- Dysmorphic features
- Myopathy
- Hepatosplenomegaly (storage disorder)
- Hepato-renal dysfunction
- Blood pressure – hypertensive cardiomyopathy may present with normal blood pressure if cardiac function is poor
- A detailed general examination and developmental assessment

Cardiomyopathy - Investigations

ECG – 12-lead and 24 hour tape required. Look for evidence of ischaemia, tachycardia, arrhythmia, low voltage complexes and repolarisation abnormalities.

Holter – rule out sustained heart rhythm problems

Chest X-ray – Assess cardiomegaly and pulmonary vascularity

Echocardiography – A full cross-sectional echocardiogram and Doppler study should be performed with particular attention paid to

- Ruling out structural heart disease e.g. left heart obstruction
- Coronary artery morphology and coronary flow
- Systolic and diastolic ventricular function (see section 5.10.3)
    - M-mode (LV study, MAPSE, TAPSE)

    - Tissue Doppler
    - Mitral regurgitation dP/dT
- Assessment of pulmonary artery pressure.
- Also look for intracardiac thrombus (in LA, LA appendage and adherent to LV wall).

Laboratory investigations:
Biochemical - Urea, electrolytes, creatinine, bone profile (calcium, phosphate and albumin and alk phos) and liver function tests (bilirubin, albumin, alk phos and ALT), CRP, cholesterol, triglycerides, urate and glucose, serum ammonia, free carnitine, acyl carnitine profile, plasma amino acids, creatinine kinase, troponin, pro-NT BNP, TFTs, autoantibodies (ANA and myocardial antibodies, need to ask for ANCA specifically if required).

Blood Gas – lactate, bicarbonate, chloride, anion gap

Genetics – in all cases, send DNA for banking. Discuss with Genetics team – consider R135 panel (paediatric cardiomyopathy)

Haematology – FBC, ESR, coagulation screen, ferritin, blood film to look for vacuolated lymphocytes.

Virology – throat or nose swab for viruses. **Request "full respiratory screen and enterovirus."**

Metabolic
Consider need for blood for alpha-glucosidase (if any features of Pompe's disease).
Consider Barth syndrome (3-Methylglutaconic aciduria type II). Think of this diagnosis in

- Males with cardiomyopathy [dilated or hypertrophic, possibly with left ventricular non-compaction and/or endocardial fibroelastosis]
- Neutropaenia [chronic, cyclic, or intermittent]
- Underdeveloped skeletal musculature and muscle weakness
- Growth delay
- Exercise intolerance
- Cardiolipin abnormalities
- Possible FHx of stillbirth.

Urine specimens - Organic acids, mucopolysaccharide screen (GAGs) in one container. Ketones, glucose and protein on dipstick.
VMA (special bottle may be required – liaise with Biochem).

Approximately **20 mls of blood** are needed (it includes ESR-4mls in EDTA), one **capillary gas** and **BM capillary** for blood sugar. However this also includes 3 mL for Barth and 3 mL for alpha-glucosidase which may not be appropriate for everyone.

Requests:
Haematology form
FBC plus blood film (d/w lab – looking for vacuolated wbc) – 1 mL EDTA
ESR – 4 mL EDTA (adult bottle not paediatric bottle)
Clotting – 1.3 paeds citrate (blue top) sample
Ferritin 1 mL plain top

Immunology test on biochemistry form
Autoantibodies, ANA and myocardial antibodies in 1 mL in plain top

1st Virology form

Throat swab or nose swab for “full respiratory screen and enteroviruses”.

2nd Virology form

Stool sample for enteroviruses

Biochemistry

5 lithium heparin samples
3 lithium heparin samples for standard biochemistry (see list below)
Ammonia assay 0.5 mL Lithium heparin sample in ICE – send within 10 minutes
0.5mL lithium heparin for plasma amino acids

Biochemistry special (if clinically indicated)

Alpha-glucosidase 3mL EDTA
Cardiolipin analysis for Barth syndrome 3mL EDTA sent to Bristol. Form to be completed.

Genetics

1-3 mL EDTA – ask for R135 panel

Urine tests Request nursing staff to send (don’t forget to give request forms)

Further specific investigations may be required for certain conditions like mitochondrial disease. Certain children with isolated hypertrophic cardiomyopathy and suspected respiratory chain disorders may require skeletal muscle biopsy, endomyocardial biopsy, DNA studies and mitochondrial respiratory chain studies. **Consider need for HIV test based on clinical presentation and also history of foreign travel and demographics.**

Cardiomyopathy Screen - Investigations

Name: Unit Number: DOB:

Genetics ☐ DNA banked _____ date ☐ R135 panel discussed on on-call Genetics team _____ date

Biochemistry Lab:

Test (units)	Date	Result	Comments
Urea		(mmol/L)	Lithium Heparin (LH)
Creatinine		(mmol/L)	LH
Sodium		(mmol/L)	LH
Potassium		(mmol/L)	LH
Bicarbonate		(mmol/L)	LH (to calculate anion gap)
Chloride		(mmol/L)	LH (to calculate anion gap)
Total Protein		(g/L)	LH
Albumin		(g/L)	LH
ALP		(IU/L)	LH
ALT		(IU/L)	LH
Bilirubin		(μmol/L)	LH
Calcium (add vit D if Ca low)		(mmol/L)	LH
Phosphate		(mmol/L)	LH
CRP		(mg/L)	LH
CK		(IU/L)	LH
Cholesterol			LH
Triglyceride			LH
Urate		(micromol/L)	LH
Troponin		(μg/L)	LH
Ammonia		(μmol/L)	LH 0.5 mL (ice sample – send within 10 min)
Free Carnitine		(μmol/L) (μmol/L)	LH
Acyl Carnitine Profile		(μmol/L)	LH
Plasma Amino acids		(μmol/L)	LH (0.5 mL)
Blood gas			Capillary
Anion gap			Calculate make sure requesting HCO_3 and Chloride in Biochemistry sample
Glucose		(mmol/L)	BM Capillary or oxalate (grey top) 0.5 mL
Lactate		(mmol/L)	Free flowing capillary blood gas tube

Thyroid Function tests		Free T4 TSH	LH
Immunology test ANA Myocardial antibodies		(goes to biochemistry lab)	1mL plain top

Haematology Lab

WCC			EDTA
Hb			
Platelet			
Blood Film			
ESR			EDTA 4mL (adult)
APTT			1.3 mL Citrate (blue top)
PT			
Fibrinogen			
Ferritin			Plain top 1 mL

Urine tests

Urine organic acids			5mL universal container
Urine-Mucopolysaccharide screen			
Urine Glucose			Dipstick urine
Urine Protein			
Urine Ketones			
Urine-VMA			Send urine in universal container - needs to be acidified ASAP by Special Biochemistry Lab

Virology

Test	Date	Results	Comments
Enterovirus			Stool sample
“Full respiratory screen and enterovirus”			On throat or nose swab (red swab –viral medium)
Additional viruses to be considered in dilated cardiomyopathy; EBV, CMV, PARVO, HSV6, and HIV etc.			Please discuss with cardiology consultant and consultant virologist

Other

Genetics (EDTA sample)			R135 gene panel
ECG			Extn 43325
24 hr Holter			Suite 11
Chest Radiograph			Extn 43027
ECHO			Extn 43920

Print out table and put at front of patient record for ready reference

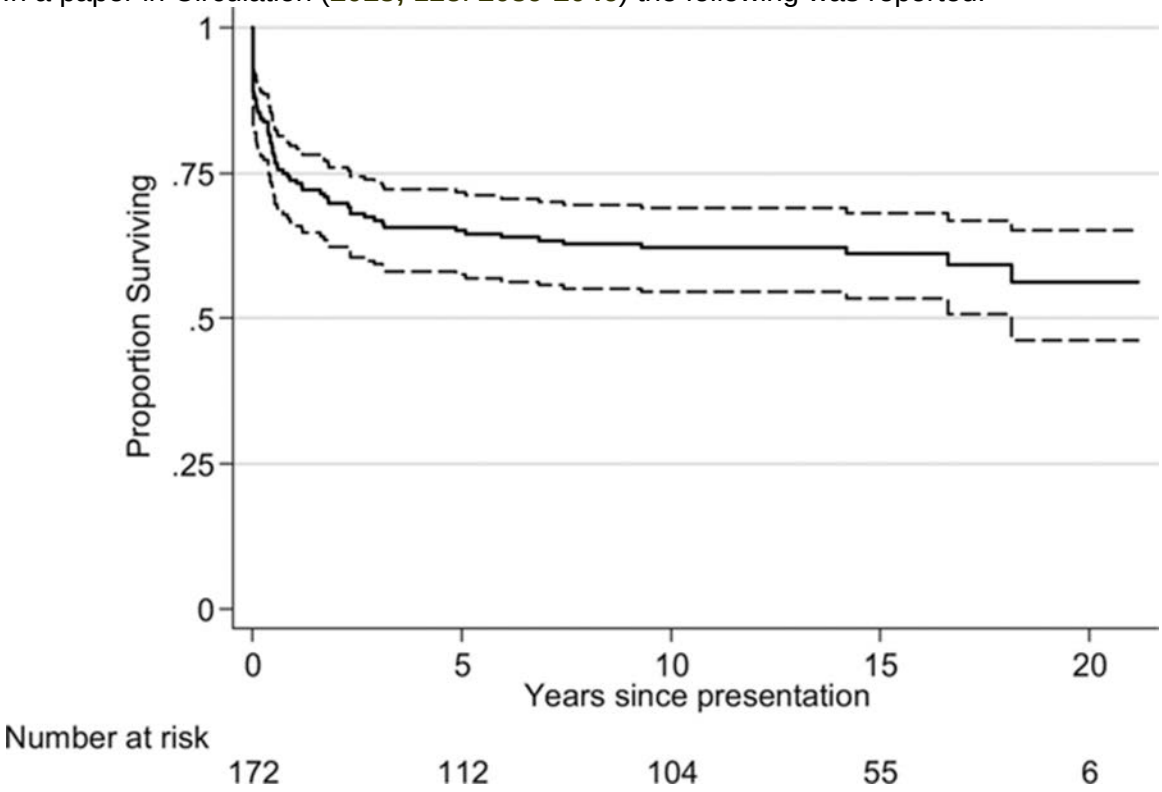
Natural History

5 year transplant-free survival following diagnosis of “cardiomyopathy” (%)

All causes	Myocarditis	Idiopathic DCM	Unclassified	Neuromuscular	Metabolic
56	78	52	70	26	33

Reference: JACC 2010;55:1377-84 – results of 189 patients, excludes pts exposed to anthracycline (small numbers)

In a paper in Circulation (**2013;** 128: **2039-2046**) the following was reported:



Worse outcomes are seen:

- In patients presenting <4/52 and >5 years of age
- When presenting ejection fraction is <20% (FS ~<10%)
- In enterovirus myocarditis
- There is intracardiac thrombus at presentation
- If severe LV dysfunction persists >3 months.
- If the DCM is familial

More favourable outcomes tend to be seen:

- In patients presenting between 1 and 5 years of age
- In those presenting with acute myocarditis (but not enterovirus)
- In patients presenting with a tachycardia-related cardiomyopathy where the rhythm can be controlled.

If LV dysfunction persists, there will eventually be a progressive deterioration with worsening cardiac failure, arrhythmias and systemic and/or pulmonary emboli.

Management – see section 6.4 above

- Diuretics

- ?Digoxin – caution in acute myocarditis – may increase risk of arrhythmia – reserve for recovery phase
- ACE inhibitors
- β-blockade – once features of acute heart failure have been stabilised (see section 3.2.3)
- Anti-platelet or anticoagulation therapy
- Cardiac transplantation – in selected cases

Screening
All forms of cardiomyopathy may be familial. Undertaking screening of family members is advised, but this may have serious consequences e.g. difficulties in obtaining life insurance or getting a mortgage. Close liaison with Medical Genetics is recommended. See Section 6.24.

See also: https://www.ahajournals.org/doi/10.1161/CIRCRESAHA.116.309386#:~:text=Within%202%20years%20of%20presentation,Sudden%20death%20is%20rare.4

Contributors: Catherine Armstrong, Victor Ofoe, Alan Pateman, Orhan Uzun, Dirk Wilson, Amos Wong
Guideline updated 2023 – due for review 2026

6.6 Chest Pain in Children

Please refer to C&V guidance – see:

General Paediatrics Clinical Portal - chest-pain-guideline-approved-july-2021.pdf - All Documents (sharepoint.com)

Most chest pain in children does not have a cardiac origin. Red flags that could indicate important chest pain, including cardiogenic pain are :

- Major Chest Trauma
- Prior cardiac disease or surgery
- Hypercoagulable states
- Sickle Cell disease
- Chronic respiratory disease
- Kawasaki disease
- Marfan syndrome or other connective tissue disorders
- Familial hyperlipidaemia syndromes
- Cocaine or stimulant use

6.7 ECMO Referral

Respiratory extra corporeal membrane oxygenation (ECMO) support is indicated for acute, severe but reversible respiratory failure when the risk of dying from the primary disease despite optimal conventional treatment is high. The duration of ECMO support is related to the underlying disease process. In general terms, it is a prolonged but temporary support (<30 days) of the lungs and sometimes the heart.

The national ECMO service allows babies and children with severe respiratory distress from a variety of causes and for whom mechanical ventilation is insufficient, to have the function of their lungs (and also heart if needed) supported with a mechanical pump and artificial lung. The causes include

- Infection
- Meconium aspiration

- Diaphragmatic hernia
- Structural problems with lungs or airway (on occasions).

ECMO provides support for gas exchange and the circulation, allowing time for intrinsic healing of damaged organs while minimising iatrogenic injury. The prerequisite for successful ECMO support is that the underlying condition is reversible.

Neonates requiring ECMO treatment need to meet the following inclusion criteria:

- Oxygenation Index * (OI) >40
- Gestational age >35 weeks
- Weight >2kgs
- Reversible lung disease

* OI = (Mean Airway Pressure [cmH20] x FiO2 [in percentage]) divided by (post ductal PaO2 [mmHg])

Exclusion Criteria

- Significant coagulopathy or uncontrollable bleeding
- Major (>grade 1) intracranial haemorrhage
- Irreversible lung injury
- Major congenital / chromosomal anomalies or severe encephalopathy
- Major cardiac malformation
- Mechanical ventilation 10 - 12 days (discussion required with ECMO team)
- Cardiac arrest other than immediately at birth

The differential diagnosis of neonatal respiratory failure and PPHN includes cyanotic congenital heart disease, particularly total anomalous pulmonary venous drainage. All patients being referred for ECMO should have a detailed echo by a neonatologist or paediatrician with echo expertise or a paediatric cardiologist. If there is any uncertainty, there needs to be consultant-to-consultant discussion.

References:
https://www.england.nhs.uk/wp-content/uploads/2013/06/e07-ecmo.pdf
http://www.leicestershospitals.nhs.uk/aboutus/departments-services/heart-services/ecmo/ecmo-treatment-criteria/

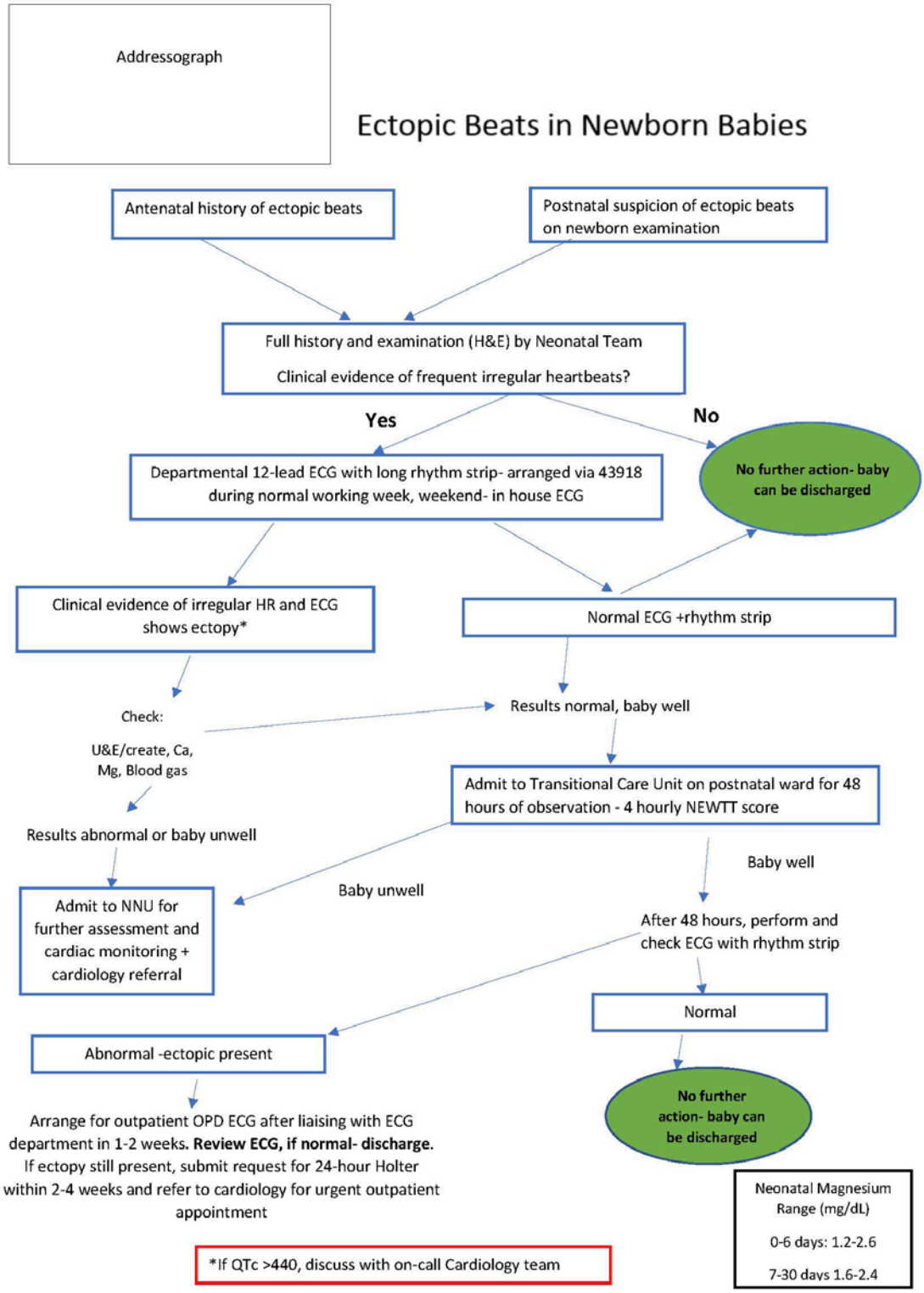
6.8 Ectopic Beats in Newborn Babies

Ectopic beats, or “extra beats” are common in newborn babies. In most cases the ectopic beats have an atrial origin and are terms premature atrial complexes (PACs) or atrial ectopic beats. In a small number of neonates the extra beats are premature ventricular complexes (PVCs).

In most cases the finding of PACs in newborn babies is not significant. They represent a developmental phenomenon and the vast majority disappear without any consequence in early infancy. A small proportion (<1-2%) may go on to develop significant arrhythmias such as SVT.
In a small proportion of cases of babies with ectopic beats there is an underlying problem with the baby and the following should be considered:

- Severe acidosis / hypoxia
- Metabolic disturbance
    - Hyperkalaemia
    - Hypocalcaemia
    - Renal failure

- Hypoglycaemia
- Infection / septicaemia
- Left atrial mass



6.9 Endocarditis

6.9.1 Infective Endocarditis

Aetiology

- Risk factors: endothelial damage + bacteraemia
- Increased risk with
    - All CHD except secundum ASD; includes most repaired defects (see 6.7.2 below)
    - Localised infection (e.g. osteomyelitis, skin infection)
    - Poor dentition or following dental/surgical procedures
    - Prosthetic heart valves
    - Indwelling vascular lines
    - Immunodeficiency, including post-transplant
    - Broken or infected skin (e.g. eczema)
- 90% due to Strep viridans, enterococci or Staphyloccoal species (aureus or coagulase-negative)

Features

- Insidious onset (usually): fatigue, fever, anorexia, pallor
- Murmur (especially changing), fever, splenomegaly, clubbing
- Petechiae, nail splinters, Osler's nodes, Janeway lesions, embolic phenomena (PE, cerebral, haematuria, Roth spots)

Investigations

- If the patient is not acutely unwell, at least three sets of blood cultures should be undertaken over the space of 12-24 hours; cultures during fever or rigor are more likely to yield positive results, but they can be taken even if the patient is afebrile. The yield of blood cultures increases with the volume of blood drawn.
- If the patient is acutely unwell take at least two sets of cultures before starting immediate treatment
- Other investigations include
    - FBC
    - U+E/creatinine
    - LFT
    - ESR/CRP
    - Coagulation profile
    - Urinalysis
    - Serial echocardiography

Treatment

The mainstay of treatment of endocarditis is antibiotic therapy. Patients with a subacute history should be treated with a combination of penicillin (4-6 weeks) and gentamicin (2 weeks). Treatment may be tailored according to the results of microbiological investigations. Patients with an acute history should additionally receive high dose flucloxacillin to cover the possibility of staphylococcal infection. If a myocardial or paravalvar abscess is shown on echocardiography, or if a vegetation is large with a high risk of embolising, early surgical discussion is required.

In staphylococcal endocarditis where vegetations are associated with prosthetic material within the heart, use of vancomycin + rifampicin may be advisable (refer to European Society of Cardiology [ESC] guidelines https://academic.oup.com/eurheartj/article/36/44/3075/2293384).

Other management points

Close consultation with the microbiologists is required.

Progress of the disease is monitored by serial FBC, ESR, CRP and echocardiography. Echocardiography may demonstrate vegetations but a negative scan does not exclude the diagnosis of endocarditis. The scan should also establish a baseline of valvular and myocardial function – both of which may change during the course of the illness. TOE is a more sensitive means of demonstrating vegetations. A dental consultation should be undertaken – it may reveal the source of infection and reduce the risk of recurrence.
Antibiotics should not be given to a stable patient until instructed by the consultant.

6.9.2 Endocarditis prophylaxis

Most cases of endocarditis do not have an identifiable precipitant, but some cases can be traced to a dental or surgical episode. NICE has published guidelines covering the prevention of IE. It is recognised that patients with CHD, prosthetic valves and post-transplant are at increased risk of IE, but prophylactic antibiotic cover should not *routinely* be prescribed to protect against infection. The NICE guideline calls for:

- Meticulous dental hygiene
- Regular dental visits
- The avoidance of body piercing and tattooing
- Prompt treatment with antibiotics of bacterial infections (e.g. abscesses or boils)
- Cessation of the practice of giving routine prophylactic antibiotics prior to certain surgical and dental procedures.

Antibiotic cover should be given for the first 6 months post intracardiac or intravascular device implant (see ECS 2015 guidance for drugs and doses). GI operations where there is a bowel abscess should be covered with antibiotics.
Reference: https://www.nice.org.uk/guidance/CG64

The ESC and AHA recommend use of antibiotic prophylaxis in certain situations in high-risk patients. ESC Guidance is summarized below:

Recommendations	Class	Level
Antibiotic prophylaxis should only be considered for patients at highest risk of IE: 1. Patients with any prosthetic valve, including a transcatheter valve, or those in whom any prosthetic material was used for cardiac valve repair. 2. Patients with previous IE. 3. Patients with congenital heart disease. a. Any cyanotic congenital heart disease. b. Any type of congenital heart disease repaired with a prosthetic material whether placed surgically or by percutaneous techniques, up to 6 months after the procedure or lifelong if residual shunt or valvular regurgitation remains.	IIa	C
Antibiotic prophylaxis is not recommended in other forms of valvular or congenital heart disease.	III	C

Procedures at highest-risk of IE

Recommendations	Class	Level
A. Dental procedures • Antibiotic prophylaxis should only be considered for dental procedures requiring manipulation of the gingival or periapical region of the teeth or perforation of the oral mucosa.	IIa	C
• Antibiotic prophylaxis is not recommended for local anaesthetic injections in non-infected tissues, treatment of superficial caries, removal of sutures, dental X-rays, placement or adjustment of removable prosthodontic or orthodontic appliances or braces, or following the shedding of deciduous teeth or trauma to the lips and oral mucosa.	III	C
B. Respiratory tract procedures • Antibiotic prophylaxis is not recommended for respiratory tract procedures, including bronchoscopy or laryngoscopy, transnasal or endotracheal intubation.	III	C
C. Gastrointestinal or urogenital procedures or TOE • Antibiotic prophylaxis is not recommended for gastroscopy, colonoscopy, cystoscopy, vaginal or caesarean delivery or TOE.	III	C
D. Skin and soft tissues procedures • Antibiotic prophylaxis is not recommended for any procedure.	III	C

If antibiotic cover is recommended, the doses are shown below:

Situation	Antibiotic	Single-dose 30–60 minutes before procedure	
		Adults	Children
No allergy to penicillin or ampicillin	Amoxicillin or Ampicillin[a]	2 g orally or i.v.	50 mg/kg orally or i.v.
Allergy to penicillin or ampicillin	Clindamycin	600 mg orally or i.v.	20 mg/kg orally or i.v.

[a]Alternatively, cephalexin 2 g i.v. for adults or 50 mg/kg i.v. for children, cefazolin or ceftriaxone 1 g i.v. for adults or 50 mg/kg i.v. for children.

Cephalosporins should not be used in patients with anaphylaxis, angio-oedema, or urticaria after intake of penicillin or ampicillin due to cross-sensitivity.

See:
https://www.ahajournals.org/doi/full/10.1161/CIR.0000000000000298 (AHA guidance)

https://academic.oup.com/eurheartj/article/36/44/3075/2293384/2015-ESC-Guidelines-for-the-management-of (ESC guidance)
and
https://www.bmj.com/content/358/bmj.j3942/rr-1

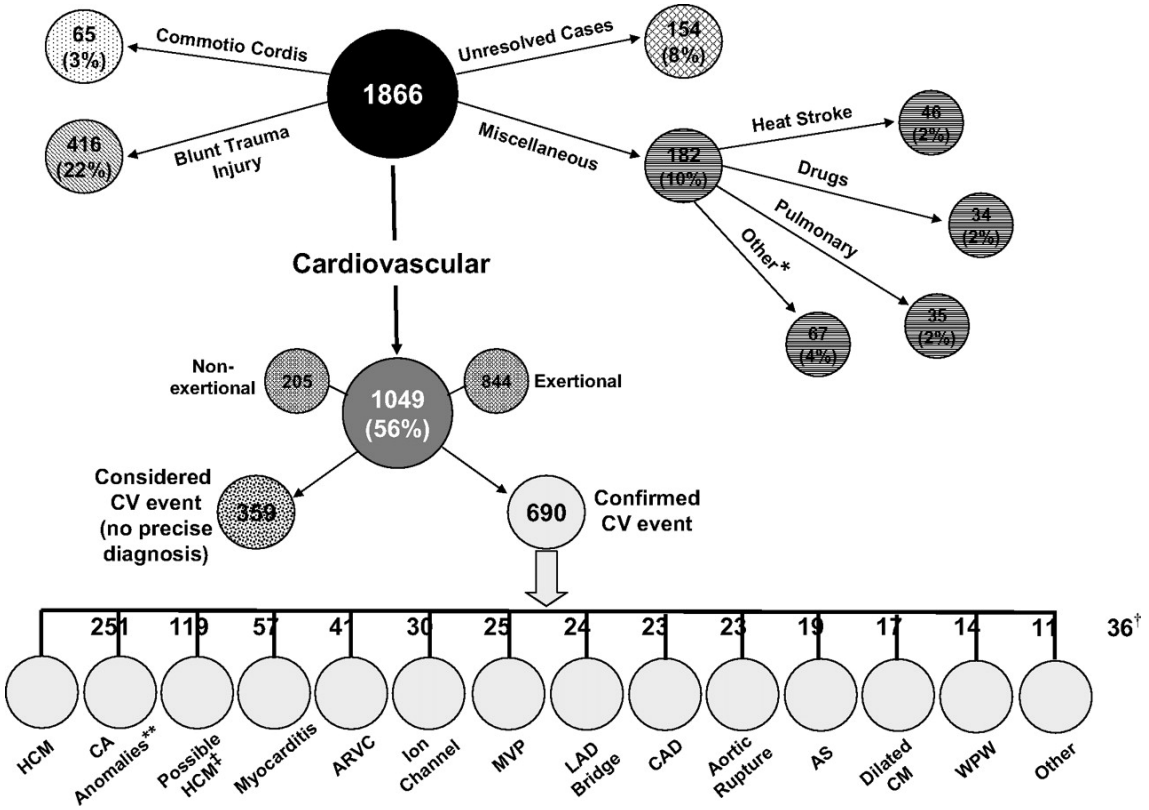
6.10 Exercise in Paediatric Cardiac Patients

Regular exercise has proven health benefits, including improved longevity, reduced obesity and better long-term cardiorespiratory performance. Children and young people with cardiac disease should participate in regular physical activity – in some there will be a need to select safe forms of activity to reduce the potential risk of harm.

See: https://www.nhs.uk/live-well/exercise/physical-activity-guidelines-children-and-young-people/

Heart Research UK has devised a Physical Activity Toolkit that allows clinicians to give specific advice on exercise type, duration and intensity. Many patients have found this toolkit to be useful. See:
https://www.swswchd.co.uk/image/Clinical%20information/Exercise/Exercise%20document%20(3).pdf

In 2009 Maron, et al, published the results of a US registry of sudden death in young athletes. The results showed (NB the numbers do not quite line up with the circles):



Sudden Deaths in Young Competitive Athletes, Circulation 2009;119(8):1085-1092

Among the 1049 cardiovascular deaths, 29% occurred in black/minority ethnic patients, 54% in high school students, and 82% with physical exertion during competition/training, whereas only 11% occurred in females (although this increased with time). In deaths where a cardiovascular rcause was firmly identified, the most common causes were hypertrophic cardiomyopathy (36%) and congenital and acquired coronary artery anomalies (23%).

General Principles

See
https://www.swswchd.co.uk/image/Clinical%20information/Exercise/Exercise%20document%20(3).pdf

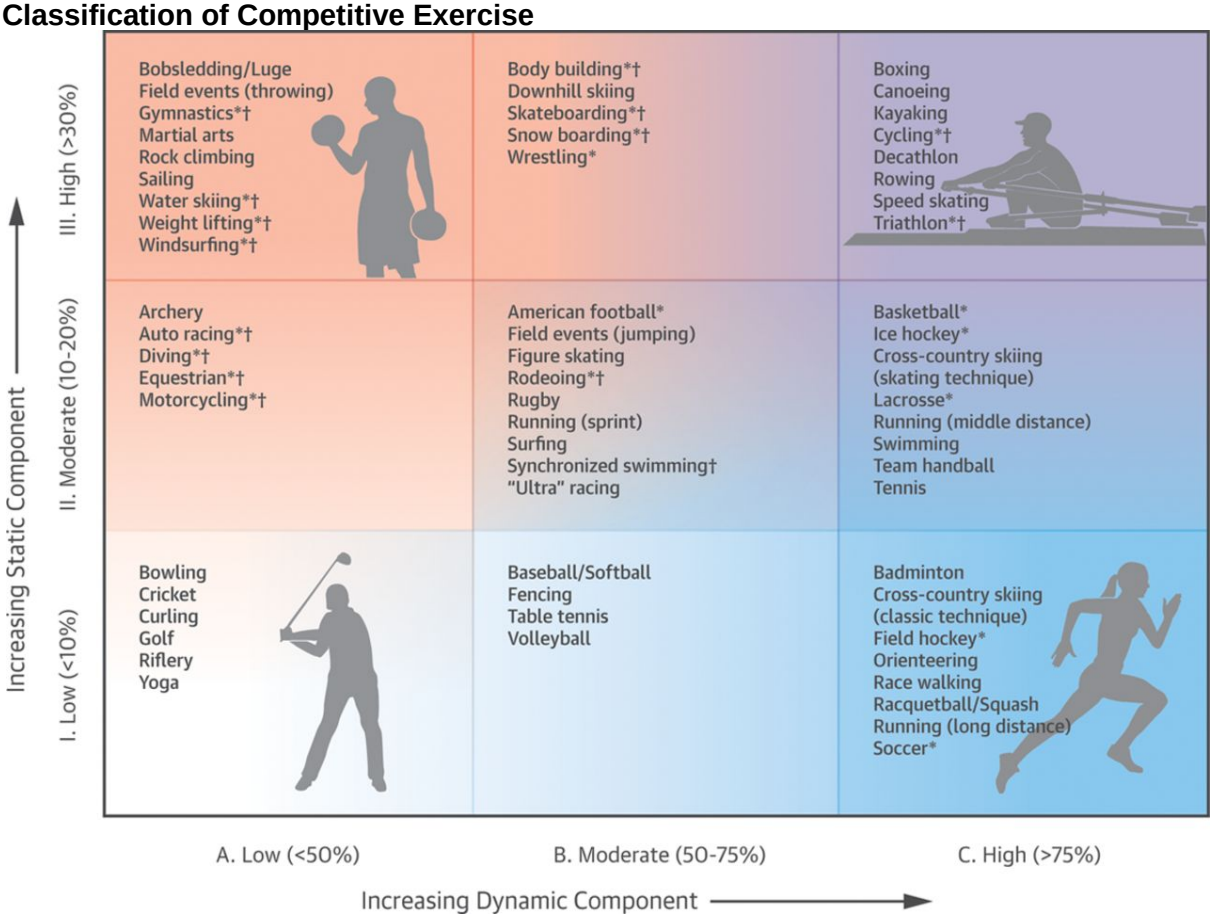
In many paediatric cardiac conditions, exercise is unrestricted, however in a number of conditions, intense exertion and competitive sports represent a risk to the health of the patient and should be restricted. The following meant to be a simple, practical guide with which clinicians can advise patients and their parents.

Although hypertrophic cardiomyopathy (HCM) is the commonest cause of sudden death in young athletes, most deaths in HCM occur with a history of minimal or no exertion. In contrast, sudden death in aortic stenosis is almost exclusively associated with exertion. Most available data and therefore the clearest recommendations relate to **athletes undertaking competitive sports** rather than the type of activities that most children engage in. It is important, therefore, to have a sense of perspective in giving advice about sports participation. Incorrect or inappropriate advice, however, may contribute to the premature death of a young person.

In general the following principles should apply:

- Patients with known HCM, severe AS, DCM, ARVC, CVPT, congenital coronary artery anomalies, enlarged aorta and LQTS should avoid high intensity/explosive sports.
- Patients with known HCM, severe AS, DCM, ARVC, CVPT, congenital coronary artery anomalies, LQTS and WPW should avoid unaccompanied cross-country running and swimming.
- Patients with LQTS, particularly LQTS type 1, should avoid diving into cold water.
- Patients with a significantly enlarged aorta (e.g. Marfan syndrome, bicuspid aortic valve spectrum, some post-op CoA, arterial switch, tetralogy and Ross patients) should not engage in high-impact contact sports or those that involve intense straining, such as rugby scrummaging, weight-lifting and rowing.
- Patients on anticoagulants or anti-platelet agents should avoid high-impact contact sports.
- Patients with exercise-induced SVT should avoid the activities that precipitate symptoms, or should take prophylactic anti-arrhythmics prior to sports participation.
- Patients with VT or high risk of VT should avoid competitive sports.
- In many cases, with appropriate advice, the school PE Department may be able to modify activities to permit at least some sports participation for the vast majority of patients.
- Patients with a pacemaker or ICD device should not engage in contact sports (risk of damage to device or electrodes). Refer to lesion-specific recommendations related to the underlying pathology for more detailed advice. See also this reference which reports good outcomes in athletes with ICD continuing with competitive sports: https://academic.oup.com/eurjpc/article/26/7/764/5925095

Classification of Competitive Exercise



This classification is based on peak static and dynamic components achieved during competition; however, higher values may be reached during training.
The increasing dynamic component is defined in terms of the estimated percentage of maximal oxygen uptake ($\dot{V}O_{2max}$) achieved and results in an increasing cardiac output. The increasing static component is related to the estimated percentage of maximal voluntary contraction reached and results in an increasing blood pressure load. The lowest total cardiovascular demands (cardiac output and blood pressure) are shown in the palest color, with increasing dynamic load depicted by increasing blue intensity and increasing static load by increasing red intensity. Note the graded transition between categories, which should be individualised on the basis of player position and style of play.
***Danger of bodily collision – avoid or advise extra precautions in patients on anti-thrombotic treatment**
†Increased risk if syncope occurs.
Modified from Mitchell et al, copyright © 2005, *Journal of the American College of Cardiology*.

Exercise advice by lesion
NB – These recommendations are for **competitive sports** during participation. Higher levels of both static and dynamic components may be attained during training. The guidelines may or may not apply to non-competitive sports participation and this is where the discretion of the clinician is needed.

For disease-specific guidance, refer to European guidance – see https://academic.oup.com/eurheartj/article/42/1/17/5898937?login=false
or American guidance – see https://www.ahajournals.org/doi/10.1161/CIR.0000000000000240#:~:text=We%20strongly%20caution%20against%20participation,testing%20or%20abnormal%20hemodynamic%20assessment.

6.11 Fetal Cardiology

The fetal cardiology service is led by Dr Pateman with support from Professor Uzun, Dr Gillett and the Fetal Cardiac Nurse Specialst, Alison Pearce. Referral into the service is via Fetal Medicine – see:
CAV Obstetrics & Gynaecology Guidelines - Fetal Medicine - Guidelines (sharepoint.com)

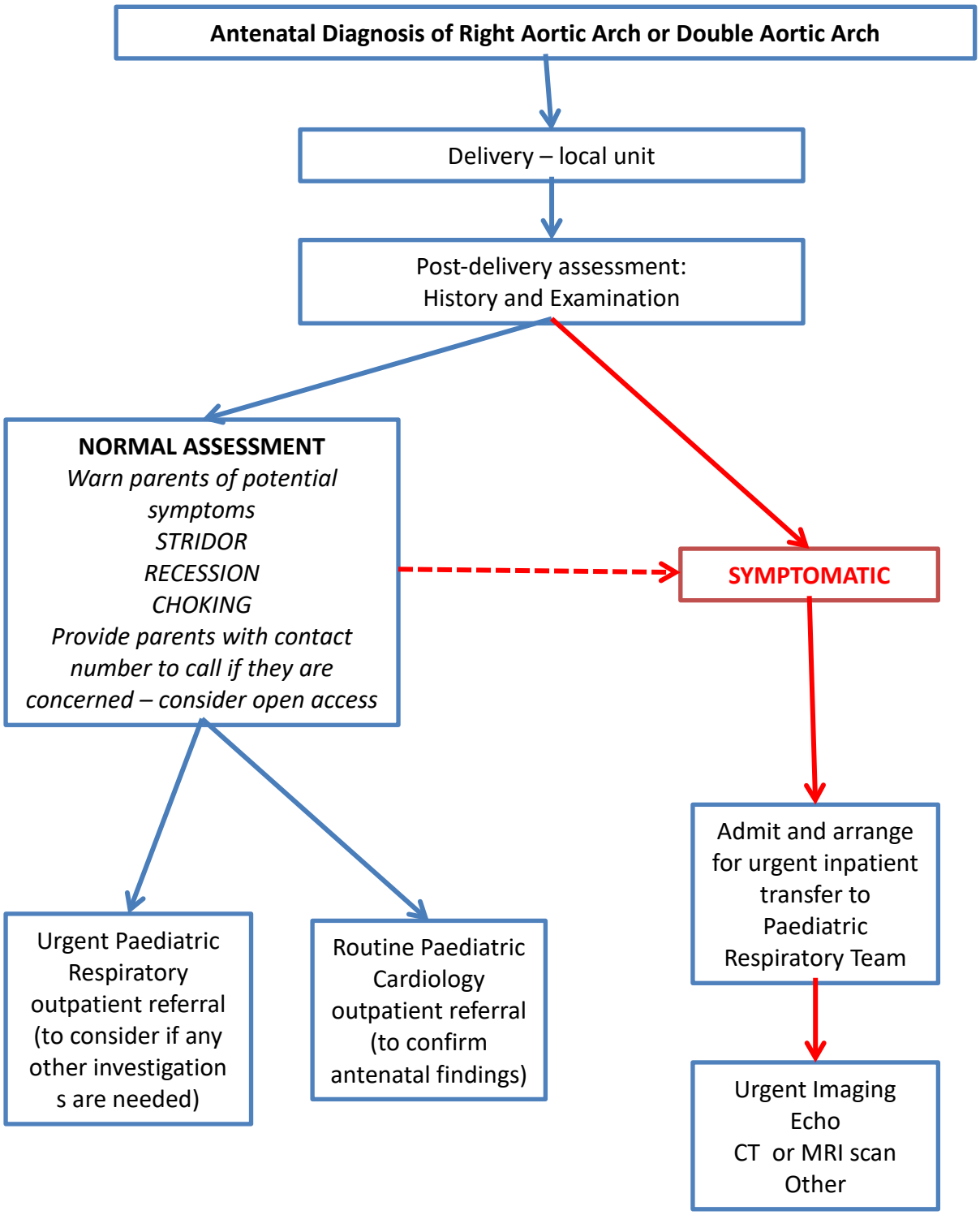
Acute obstetric services have the responsibility to deal with perceived cardiac emergencies in fetuses, supported by fetal medicine and paediatric/fetal cardiology during working hours. Generic paediatric cardiology advice is available from the on-call paediatric cardiologist, but there is no expectation than non-experts would undertake hands-on assessment.

6.11.1 Fetal SVT

Please refer to the UHB guidance set out below:
CAV Obstetrics & Gynaecology Guidelines - Fetal Heart Irregularities and Arrhythmias.pdf - Guidelines (sharepoint.com)

6.11.2 Post-natal management of antenatally diagnosed aortic arch anomalies

Examples include right aortic arch and double aortic arch.



6.12 Fainting and Syncope

Definition of syncope:

"Transient loss of consciousness (T-LOC) due to transient global cerebral hypoperfusion characterised by rapid onset, short duration and spontaneous complete recovery"

See:
https://www.ncbi.nlm.nih.gov/pmc/articles/PMC7469873/#:~:text=In%20summary%2C%20syncope%20is%20a,because%20of%20serious%20medical%20conditions.
And
https://academic.oup.com/eurheartj/article/39/21/1883/4939241?login=false

For POTS advice, see: https://www.onlinecjc.ca/article/S0828-282X(19)31550-8/fulltext

Red flags

Refer/discuss urgently for specialist cardiovascular assessment by Paediatrician with Cardiology Interest or Paediatric Cardiology (UHW), anyone with TLoC who also has any of the following.

- An ECG abnormality (see above)
- Heart failure (history or physical signs)
- TLoC *during* exertion
- Family history of sudden cardiac death in people aged younger than 40 years and/or an inherited cardiac condition
- New or unexplained breathlessness
- A heart murmur (unless clearly a flow murmur) and /or abnormal femoral pulses

Cardiac syncope

Refer/discuss urgently for specialist cardiovascular assessment by Paediatrician with Cardiology Interest or Paediatric Cardiology (UHW).

For individuals who have experienced syncope **during exercise:**

Advise the person to refrain from exercise until informed otherwise following further assessment

Tilt testing may be valuable in the following situations:

and exercise testing is not available locally and requires referral to Paediatric Cardiology at UHW.

This is usually arranged following assessment by local Paediatrician.

Treatment

Usually reassurance and information on the causes and the benign nature of most syncope is sufficient. The departmental information leaflet can be useful (see Z:\PaedCard\SHARED\INFO SHEETS\VASOVAGAL-SYNCOPE

Consider the value of undertaking a **tilt test** – this may be advisable if symptoms are atypical or if drug treatment is being considered.

Drug Treatment (specialist advice needed)

- *Salt* supplementation may be used in selected patients with no contraindications: the few studies available in small numbers of young patients used 120 mmol of salt (as slow sodium, 12 tablets per day in divided doses) daily in patients with 24-hour urinary sodium estimations of <170 mmol/24 hour. Patients are unlikely to tolerate more than 3–4 tablets twice daily because of nausea and vomiting. Blood pressure should be monitored closely, with discontinuation of salt therapy attempted after 1 year.

- *Fludrocortisone:* 50 µg once daily for 1 week, if tolerated increasing to 100 µg once daily and reviewed after 1 month. The maximum dose is 300 µg once daily. Supine blood pressure monitoring and 4–6 monthly electrolyte monitoring are mandatory.
- *Midodrine:* This α-adrenoceptor agonist has no UK license, but can be made available on a named patient basis. See section 7.13.
- *Ivabradine:* This can be considered in patients with a POTS picture overlapping with vasovagal syncope. See section 7.11.

Permanent pacing in vasovagal syncope

Evidence for the efficacy of permanent pacing in the management of vasovagal syncope is contradictory. Pacing may be considered in rare cases of "malignant" vasovagal syncope where there are repeated, unheralded, often injurious vasovagal syncope, with prolonged asystole on tilt testing, >3 seconds asystole during real-time syncope with pauses (on the table or with a Reveal device). Dual chamber pacing is mandatory, preferably DDI with hysteresis or a specifically designed algorithm for neurally mediated disorders (for example, rate drop response [for example, Adapta DR, Medtronic Inc), closed loop stimulation (for example, Cylos CLS, Biotronik]).

Useful information websites for families

www.stars.org.uk and www.nice.org.uk/guidance/cg109 (applies to teenagers only)

Contributors: Marion Schmidt, Dirk Wilson – due for review 2026

6.13 Home Monitoring Programme for High Risk Patients

Objectives

To reduce adverse events and mortality in high risk cardiac patients.

Eligibility

The HMP will be utilised in every infant with

- The Norwood operation or hybrid procedure for HLHS
- A Blalock-Taussig shunt
- Other patients deemed to be at “high risk” of an adverse event.

Pre-Discharge Criteria

- There has been acceptable weight gain for the past 24-48 hours
- Saturations in room air are within the acceptable range of 75-89%
    1. If ≤70%, the echocardiogram has demonstrated satisfactory shunt appearance on 2-D and colour flow; the branch pulmonary arteries look satisfactory
    2. If ≤70% because of long-term pulmonary pathology, this is being treated and home oxygen has been arranged (NB sort-term lung problems should be resolved before discharge)
    3. If ≥90% the patient is on appropriate diuretics and an ACE inhibitor; ventricular function is satisfactory on echocardiography
- Any active general medical problems have been addressed; any diarrhoea or vomiting has settled; if there has been weight loss there is no clinical or biochemical evidence of dehydration (including satisfactory U&E/creatinine)

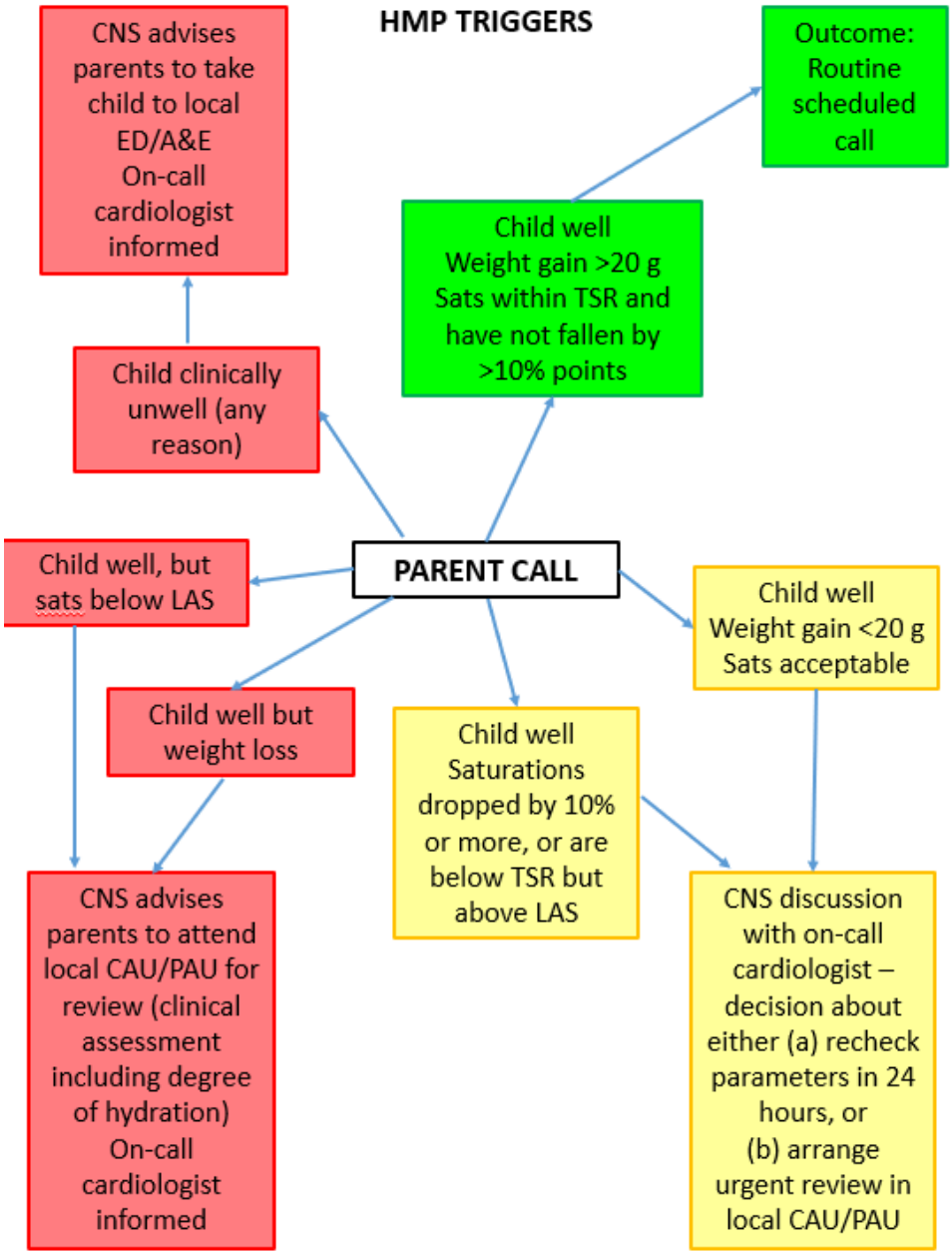
Process

- All pre-discharge criteria are met
- Parental training:
    - Principles of the HMP

    - Feeding and medication regimen
    - Understanding of “call criteria” (escalation criteria)
    - Ability to use pulse oximeter
    - Understanding of home oxygen system (if relevant)
- Patients discharged home with user-friendly baby weighing scales, oxygen saturation monitors and patient documentation containing the “call criteria”
- Documentation
    - Call criteria (“triggers”)
    - Notification of contact names and numbers.
    - Medication, nutritional feeding plan and a table of HMP parameters:
        - Weight
        - SaO_2
        - Respiratory rate
        - Heart rate
        - Feed intake
- If the child is still on the HMP at 6 months the frequency of recordings can be reduced to once weekly at the discretion of the Consultant in charge.
- Parents or community nurses will contact the Children’s Heart Nurse Specialist by telephone on the day of review and report the baby’s clinical status and HMP parameters
- The Children’s Heart Nurse Specialists will review trends and consider any problems
- An electronic copy of HMP parameters will be held by the Children’s Heart Nurse Specialists in the dedicated spreadsheet on the shared drive
- Parents are instructed to report any changes in parameters or if they have any questions or concerns
- The purpose of this monitoring is not to deal with emergency situations. **If the patient acutely deteriorates at home parents should dial 999 to contact Emergency Services for urgent admission via A+E.**

Documentation
HMP parameters will be documented on an Excel spreadsheet stored in the Home Monitoring Folder of the shared drive. All parental calls for breach of criteria or general questions will be documented on Cardiobase.

HMP Escalation Criteria (“HMP triggers”) – see HMP flow chart below



Each child has
(1) Target saturation range (TSR), e.g. 75-85%
(2) Lowest acceptable saturation (LAS), e.g. 70%

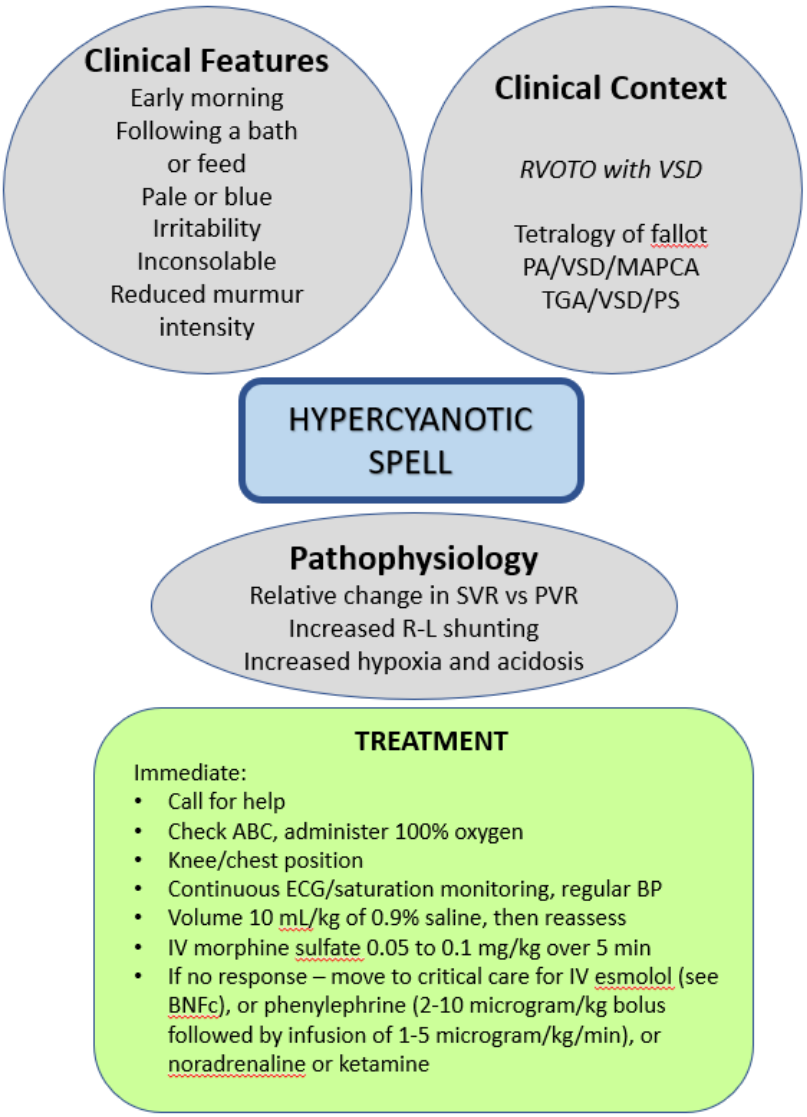
Reference

Ghanayem et al. Home monitoring program of infants after stage 1 palliation of hypoplastic left heart syndrome. *Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu* 2004;7:32-38

Contributors: Chris Gillett, Alison Pearce, Kayla Price, Asmita Raja, Dirk Wilson, Amos Wong
Due for review 2026

6.14 Hypercyanotic Spells

IF A PARENT TELEPHONES REPORTING A SIGNIFICANT SPELL ARRANGE FOR IMMEDIATE HOSPITAL ADMISSION



Long-term treatment – consider

- Prophylactic long-term propranolol
- Surgical shunt (may be needed urgently)
- Pulmonary valvuloplasty if obstruction is predominately valvular
- Transcatheter stenting of RV outflow tract, or ductus arteriosus if still present
- Early surgical repair

Hypercyanotic episodes may also occur in patients with Eisenmenger syndrome due to a sudden increase in pulmonary vascular resistance or drop in systemic vascular resistance. Give oxygen, volume expansion and consider use of morphine. Consider the need for targeted pulmonary hypertension therapy and balloon atrial septostomy or Potts shunt.

Parents of children with unrepaired tetralogy of Fallot should be given the unit information leaflet on spelling.

See also https://www.swswchd.co.uk/en/page/clinical-information-children and click on “Spell” link.

Contributor: Dirk Wilson

6.15 Hypertension in Children

Hypertension is defined as a blood pressure >95th percentile for age and height. Paediatric hypertension, over many years, will be a cause of significant morbidity and mortality and it is being increasingly recognised. Paediatric cardiologists have a role in the management of hypertension: (a) to rule out aortic obstruction as the cause, and (b) to assess end-organ involvement, specifically using echo to note changes in LV wall thickness, mass and function.

A significant proportion of patients with treated coarctation have hypertension – this may be immediately apparent after surgery, but may also arise many months or years after treatment. It has been noted that paediatric cardiologists are not always attentive at diagnosing and treating post-coarctation hypertension.

Ambulatory blood pressure assessment plays an important role in the diagnosis and medical management of childhood hypertension (see section 5.6).

The incidence of hypertension has been assessed in a UK study:
https://www.nature.com/articles/s41371-022-00732-7

UK dynamap-derived age-based systolic and diastolic BP centiles can be found through this link: 298.pdf (nih.gov)

A paediatric blood pressure calculator can be found at this link:
https://hyperchildnet.eu/blood-pressure-calculator/

The European Society of Hypertension has published guidance on the diagnosis, investigation and management of hypertension in children:
https://www.eshonline.org/esh-content/uploads/2021/10/2016-European-Society-of-Hypertension-guidelines-for-the-management-of-high-blood-pressure-in-children-and-adolescents.pdf

Age/Sex based BP centile charts of European children are shown below:

Age-based BP centiles for BOYS:

		Systolic (mmHg) percentile of height							Diastolic (mmHg) percentile of height						
Age (years)	BP percentile	5th	10th	25th	50th	75th	90th	95th	5th	10th	25th	50th	75th	90th	95th
1	90th	94	95	97	99	100	102	103	49	50	51	52	53	53	54
	95th	98	99	101	103	104	106	106	54	54	55	56	57	58	58
	99th	105	106	108	110	112	113	114	61	62	63	64	65	66	66
2	90th	97	99	100	102	104	105	106	54	55	56	57	58	58	59
	95th	101	102	104	106	108	109	110	59	59	60	61	62	63	63
	99th	109	110	111	113	115	117	117	66	67	68	69	70	71	71
3	90th	100	101	103	105	107	108	109	59	59	60	61	62	63	63
	95th	104	105	107	109	110	112	113	63	63	64	65	66	67	67
	99th	111	112	114	116	118	119	120	71	71	72	73	74	75	75
4	90th	102	103	105	107	109	110	111	62	63	64	65	66	66	67
	95th	106	107	109	111	112	114	115	66	67	68	69	70	71	71
	99th	113	114	116	118	120	121	122	74	75	76	77	78	78	79
5	90th	104	105	106	108	110	111	112	65	66	67	68	69	69	70
	95th	108	109	110	112	114	115	116	69	70	71	72	73	74	74
	99th	115	116	118	120	121	123	123	77	78	79	80	81	81	82
6	90th	105	106	108	110	111	113	113	68	68	69	70	71	72	72
	95th	109	110	112	114	115	117	117	72	72	73	74	75	76	76
	99th	116	117	119	121	123	124	125	80	80	81	82	83	84	84
7	90th	106	107	109	111	113	114	115	70	70	71	72	73	74	74
	95th	110	111	113	115	117	118	119	74	74	75	76	77	78	78
	99th	117	118	120	122	124	125	126	82	82	83	84	85	86	86
8	90th	107	109	110	112	114	115	116	71	72	72	73	74	75	76
	95th	111	112	114	116	118	119	120	75	76	77	78	79	79	80
	99th	119	120	122	123	125	127	127	83	84	85	86	87	87	88
9	90th	109	110	112	114	115	117	118	72	73	74	75	76	76	77
	95th	113	114	116	118	119	121	121	76	77	78	79	80	81	81
	99th	120	121	123	125	127	128	129	84	85	86	87	88	88	89
10	90th	111	112	114	115	117	119	119	73	73	74	75	76	77	78
	95th	115	116	117	119	121	122	123	77	78	79	80	81	81	82
	99th	122	123	125	127	128	130	130	85	86	86	88	88	89	90
11	90th	113	114	115	117	119	120	121	74	74	75	76	77	78	78
	95th	117	118	119	121	123	124	125	78	78	79	80	81	82	82
	99th	124	125	127	129	130	132	132	86	86	87	88	89	90	90
12	90th	115	116	118	120	121	123	123	74	75	75	76	77	78	79
	95th	119	120	122	123	125	127	127	78	79	80	81	82	82	83
	99th	126	127	129	131	133	134	135	86	87	88	89	90	90	91
13	90th	117	118	120	122	124	125	126	75	75	76	77	78	79	79
	95th	121	122	124	126	128	129	130	79	79	80	81	82	83	83
	99th	128	130	131	133	135	136	137	87	87	88	89	90	91	91
14	90th	120	121	123	125	126	128	128	75	76	77	78	79	79	80
	95th	124	125	127	128	130	132	132	80	80	81	82	83	84	84
	99th	131	132	134	136	138	139	140	87	88	89	90	91	92	92
15	90th	122	124	125	127	129	130	131	76	77	78	79	80	80	81
	95th	126	127	129	131	133	134	135	81	81	82	83	84	85	85
	99th	134	135	136	138	140	142	142	88	89	90	91	92	93	93
16	90th	125	126	128	130	131	133	134	78	78	79	80	81	82	82
	95th	129	130	132	134	135	137	137	82	83	83	84	85	86	87
	99th	136	137	139	141	143	144	145	90	90	91	92	93	94	94
17	90th	127	128	130	132	134	135	136	80	80	81	82	83	84	84
	95th	131	132	134	136	138	139	140	84	85	86	87	87	88	89
	99th	139	140	141	143	145	146	147	92	93	93	94	95	96	97

BP, blood pressure. Modified from Task Force on High Blood Pressure in Children and Adolescents [24].

Age based BP centiles for GIRLS:

		Systolic (mmHg) percentile of height							Diastolic (mmHg) percentile of height						
Age (years)	BP percentile	5th	10th	25th	50th	75th	90th	95th	5th	10th	25th	50th	75th	90th	95th
1	90th	97	97	98	100	101	102	103	52	53	53	54	55	55	56
	95th	100	101	102	104	105	106	107	56	57	57	58	59	59	60
	99th	108	108	109	111	112	113	114	64	64	65	65	66	67	67
2	90th	98	99	100	101	103	104	105	57	58	58	59	60	61	61
	95th	102	103	104	105	107	108	109	61	62	62	63	64	65	65
	99th	109	110	111	112	114	115	116	69	69	70	70	71	72	72
3	90th	100	100	102	103	104	106	106	61	62	62	63	64	64	65
	95th	104	104	105	107	108	109	110	65	66	66	67	68	68	69
	99th	111	111	113	114	115	116	117	73	73	74	74	75	76	76
4	90th	101	102	103	104	106	107	108	64	64	65	66	67	67	68
	95th	105	106	107	108	110	111	112	68	68	69	70	71	71	72
	99th	112	113	114	115	117	118	119	76	76	76	77	78	79	79
5	90th	103	103	105	106	107	109	109	66	67	67	68	69	69	70
	95th	107	107	108	110	111	112	113	70	71	71	72	73	73	74
	99th	114	114	116	117	118	120	120	78	78	79	79	80	81	81
6	90th	104	105	106	108	109	110	111	68	68	69	70	70	71	72
	95th	108	109	110	111	113	114	115	72	72	73	74	74	75	76
	99th	115	116	117	119	120	121	122	80	80	80	81	82	83	83
7	90th	106	107	108	109	111	112	113	69	70	70	71	72	72	73
	95th	110	111	112	113	115	116	116	73	74	74	75	76	76	77
	99th	117	118	119	120	122	123	124	81	81	82	82	83	84	84
8	90th	108	109	110	111	113	114	114	71	71	71	72	73	74	74
	95th	112	112	114	115	116	118	118	75	75	75	76	77	78	78
	99th	119	120	121	122	123	125	125	82	82	83	83	84	85	86
9	90th	110	110	112	113	114	116	116	72	72	72	73	74	75	75
	95th	114	114	115	117	118	119	120	76	76	76	77	78	79	79
	99th	121	121	123	124	125	127	127	83	83	84	84	85	86	87
10	90th	112	112	114	115	116	118	118	73	73	73	74	75	76	76
	95th	116	116	117	119	120	121	122	77	77	77	78	79	80	80
	99th	123	123	125	126	127	129	129	84	84	85	86	86	87	88
11	90th	114	114	116	117	118	119	120	74	74	74	75	76	77	77
	95th	118	118	119	121	122	123	124	78	78	78	79	80	81	81
	99th	125	125	126	128	129	130	131	85	85	86	87	87	88	89
12	90th	116	116	117	119	120	121	122	75	75	75	76	77	78	78
	95th	119	120	121	123	124	125	126	79	79	79	80	81	82	82
	99th	127	127	128	130	131	132	133	86	86	87	88	88	89	90
13	90th	117	118	119	121	122	123	124	76	76	76	77	78	79	79
	95th	121	122	123	124	126	127	128	80	80	80	81	82	83	83
	99th	128	129	130	132	133	134	135	87	87	88	89	89	90	91
14	90th	119	120	121	122	124	125	125	77	77	77	78	79	80	80
	95th	123	123	125	126	127	129	129	81	81	81	82	83	84	84
	99th	130	131	132	133	135	136	136	88	88	89	90	90	91	92
15	90th	120	121	122	123	125	126	127	78	78	78	79	80	81	81
	95th	124	125	126	127	129	130	131	82	82	82	83	84	85	85
	99th	131	132	133	134	136	137	138	89	89	90	91	91	92	93
16	90th	121	122	123	124	126	127	128	78	78	79	80	81	81	82
	95th	125	126	127	128	130	131	132	82	82	83	84	85	85	86
	99th	132	133	134	135	137	138	139	90	90	90	91	92	93	93
17	90th	122	122	123	125	126	127	128	78	79	79	80	81	81	82
	95th	125	126	127	129	130	131	132	82	83	83	84	85	85	86
	99th	133	133	134	136	137	138	139	90	90	91	91	92	93	93

BP, blood pressure. Modified from Task Force on High Blood Pressure in Children and Adolescents [24].

The decision to initiate antihypertensive treatment should be based on the BP levels in combination with consideration of the presence/absence of end-organ damage, and other risk factors such as obesity, renal disease and diabetes. Management of co-existing risk factors is important, e.g. obesity, high salt intake and low physical activity. Consideration has to be given to the risks vs benefits of prolonged (life-long) drug administration. Once drug treatment is initiated, the target BP should be <95th (and ideally <90th) percentile for age/height.

Long-term oral antihypertensive agents include:

A ACE-inhibitors, angiotensin receptor blockers
B Beta-blockers
C Calcium channel blockers
D Diuretics
O Other agents (e.g. alpha-receptor blocking agents, methyldopa, minoxidil, clonidine, hydralazine)

See table below for indications/contra-indications:

Antihypertensive class	Recommended indication	Contraindicated
Diuretics	Hyperaldosteronism	Chronic renal failure
Potassium-sparing diuretics	Chronic renal failure (watch K+)	
Loop diuretics	Congestive heart failure with fluid retention	
Beta-blockers	Coarctation of the aorta Congestive heart failure	Bronchial asthma
Calcium channel blockers	Post renal transplant	Bilateral renal artery stenosis (RAS)
ACE inhibitors	Chronic kidney disease Diabetes mellitus Congestive heart failure	Bilateral RAS or RAS in single kidney Hyperkalaemia Pregnancy
Angiotensin receptor blockers	Chronic kidney disease Diabetes mellitus Congestive heart failure	Bilateral RAS or RAS in single kidney Hyperkalaemia Pregnancy
IV vasodilators	Life-threatening conditions	

Contributors: Dirk Wilson, Gareth Davies – due for review 2026

6.16 Kawasaki Disease

Diagnostic criteria
Fever for longer than 5 days plus at least 4 of:

- Non-purulent conjunctivitis
- Changes in the mouth (strawberry tongue or red, cracked lips)
- Changes in the periphery (erythema, oedema, of feet + hands)
- Polymorphous rash
- Cervical lymphadenopathy (>1.5 cm)

Additional features
Acute phase (up to10 days)

- Irritability (>90%) – may have aseptic meningitis
- Sterile pyuria (70%)
- Arthritis (40%)
- GI symptoms (25%)
- ECG changes
- Myocarditis
- Pericarditis

Sub-acute phase (>10 days)

- Desquamation
- Coronary aneurysms (10-20% if not given IVIG)
- Pericardial effusion
- Thrombocytosis

Atypical Kawasaki Disease

- May occur in some patients, more often in infants
- Full diagnostic features not seen – often just fever and 1-2 others; no other explanation for symptoms
- Should be considered in any infant with fever lasting >5 days and no other explanation
- Laboratory features are compatible
- Often complicated by coronary artery aneurysm because of delayed presentation
- Diagnosis and management are guided by echo features and lab findings

Clinical Course of KD

Acute phase (up to 10 days)

Persistent pyrexia, irritability with bilateral conjunctivitis and rash. Hands and feet develop the erythema and oedema. Tongue and oral mucosa become red and cracked. Hepatic dysfunction, myocarditis and pericarditis may develop.

Subacute stage (days 11-20)

Persistent irritability, anorexia, and conjunctival injection. Resolution of fever (if it persists the greater the risk of cardiac complications). Thrombocytosis develops. Desquamation of the fingertips and toes begins. Aneurysm formation may occur.

Convalescent phase (days 21-60)

The most significant clinical finding that persists through this phase is the presence of coronary artery aneurysms.

Chronic phase

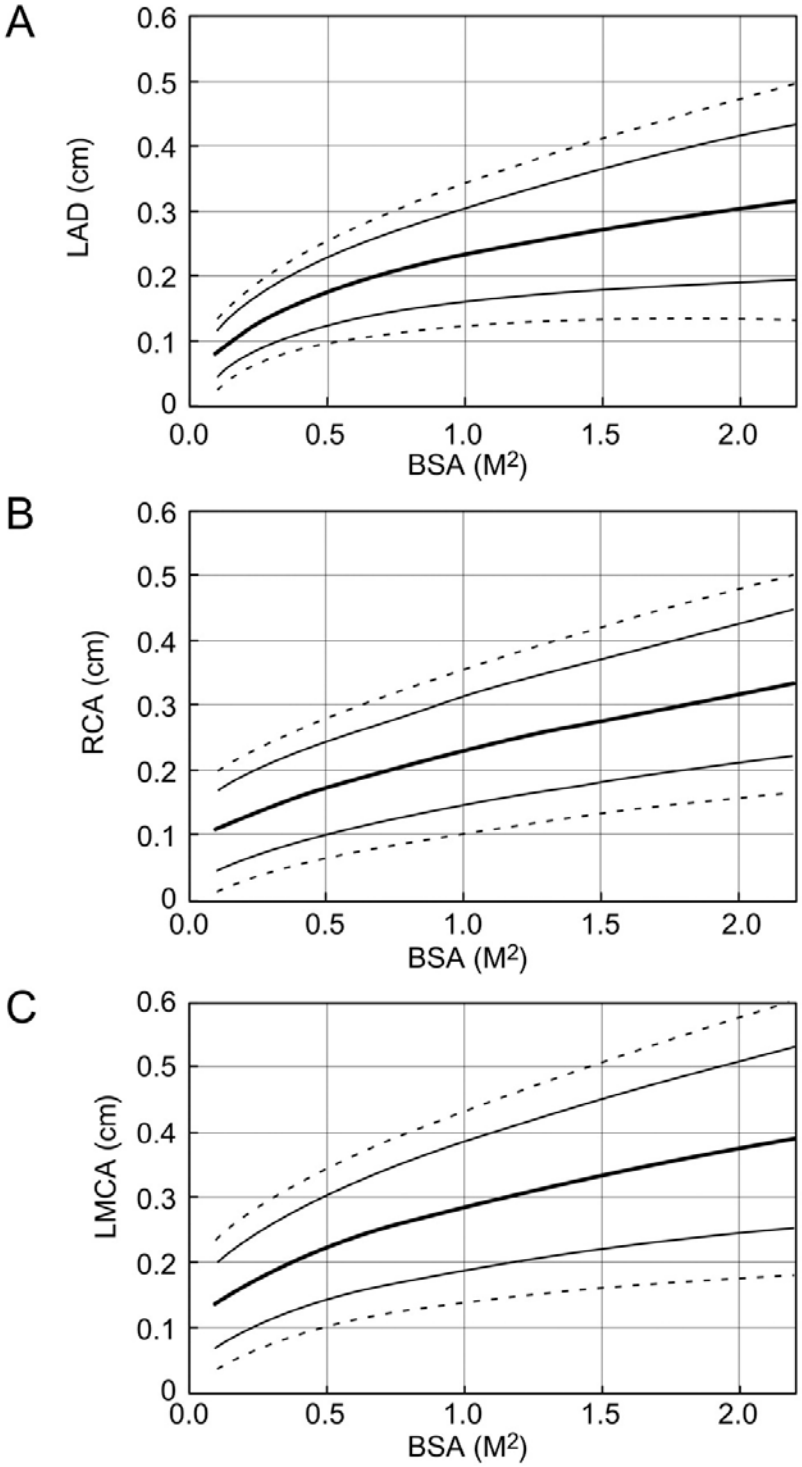
This stage is only of clinical importance in patients who have developed cardiac complications. Its duration is of lifetime significance since the aneurysm formed in childhood may thrombose or rupture in adulthood. In many cases of adults presenting with coronary artery aneurysm, careful reviews of past medical histories have revealed febrile childhood illnesses of unknown aetiology.

Investigations

ESR (often > 100)
CRP
WBC
Platelets (may be > 1000)
ASO titre to exclude streptococcal infection
There may be evidence of anaemia, pyuria and deranged LFTs
Echo features

- Coronary artery aneurysms
- Myocarditis
- Pericarditis
- Valve regurgitation
- May be normal

Coronary Artery Z-scores



Z-Score Classification of Coronaries (AHA guideline)

1. No involvement: Always <2
2. Dilation only: 2 to <2.5; or if initially <2, a decrease in Z score during follow-up ≥1
3. Small aneurysm: ≥ 2.5 to <5
4. Medium aneurysm: ≥5 to <10, and absolute dimension <8 mm
5. Large or giant aneurysm: ≥10, or absolute dimension ≥8 mm

Treatment

Acute phase (up to 10 days):

- IVIG 2g/kg single dose as slow i.v. infusion (preferred), or IVIG 400 mg/kg/day for 4 days
- Aspirin is given in an anti-inflammatory dose until
  - the child has been afebrile for 48 to 72 hours, **or**
  - the ESR/CRP normalise, **or**
  - day 14 of illness and ≥48 to 72 hours after fever cessation.
  - Recommended dose acute phase is either
    30-50 mg/kg/day in 4 divided doses (UK recommendations), **or**
    80-100 mg/kg/day given in 4 divided doses (USA and Japan recommendations).
    Then change to low-dose aspirin (3 to 5 mg/kg per day) and maintain it until the patient shows no evidence of coronary changes by 6 to 8 weeks after the onset of illness.

Low-dose aspirin should be continued for as long as coronary abnormalities persist. Avoid concomitant use of ibuprofen (antagonises the irreversible platelet inhibition induced by aspirin).

Subacute phase (>10 days): aspirin 3-5 mg/kg/day; discontinue at 6-8 weeks if no coronary involvement.

If there is no response to IVIG or recurrence of fever within a few days, reconsider the diagnosis and consider a second dose of IVIG. Further failure to respond may be treated with:

- Corticosteroids, either
  - Intravenous pulse methylprednisolone, 30 mg/kg for 2 to 3 hours, administered once daily for 3 days, with or without a subsequent course and taper of oral prednisolone, or
  - Prednisolone 2 mg/kg/day, 3 times daily, given by IV injection for 5 days or until the fever resolves and then orally 2 mg/kg/day until the C-reactive protein (CRP) level normalizes, then taper over 15 days in 5-day steps (2 mg/kg/day for 5 days, 1 mg/kg/day for 5 days, and 0.5 mg/kg/day for 5 days).
- Infliximab can be considered for resistant cases. Discuss with the Paediatric Infectious Diseases Team.

Patients presenting >10 days, but with coronary artery aneurysms should receive IVIG even if there is no good evidence of active disease.

Administration of a primary IVIG/corticosteroid combination should be considered in patients with features of the most severe disease (and therefore the greatest likelihood of developing CAA), including

- The very young (<6-12 months old)
- Those with markers of severe disease, including
  - Intense inflammation, CRP >100
  - Neutrophil count >80% of total white cells
  - Initial platelet count <30
  - Liver dysfunction, especially elevated aspartate aminotransferase
  - Hypoalbuminaemia
  - Serum sodium <133 mmol/L
  - Anaemia
  - Organ dysfunction
  - Patients that develop haemophagocytic lymphohistiocytosis (HLH).

References: *Lancet* 2012:379:1613-1620; *Arch Dis Child* 2014;99:74-83;
http://heart.bmj.com/content/early/2012/10/18/heartjnl-2012-302407.full.pdf+html
http://circ.ahajournals.org/content/110/17/2747.full

Immunisation following Kawasaki disease
Immunisation with all vaccines should be deferred at least 3 months following an episode of KD treated with IVIG. Measles, mumps, and varicella immunizations should be deferred for 11 months after receiving high dose IVIG. However, children in whom risk of exposure to measles is high may receive vaccination earlier and then be re-immunized at least 11 months after IVIG administration if they have an inadequate serological response (AHA 2017). Patients who will require long-term aspirin should be considered for Varicella zoster virus vaccination (to reduce the risk of Reye's syndrome).

Monitoring
The acute phase proteins and platelets should be monitored until normal. A baseline echocardiogram should be undertaken. If this echo is abnormal, it should be repeated as clinically indicated. If the initial echo is normal, it should be repeated at 2 weeks, then again at 6-8 weeks and 6-8 months from initial presentation.
If the 6-8 week echo is normal, consider stopping the aspirin.
If the echocardiogram and ECG are normal at six months then there is no evidence that new coronary abnormalities will be identified after this period, although the AHA guidelines suggest continuing follow-up for one year and UK guidelines suggest indefinite follow-up is needed (see table below from *Arch Dis Child* 2014;99:74-83).

All patients treated with steroids should be followed to identify adverse effects including osteonecrosis, adrenal supression and intercurrent infection.

Risk stratification and follow-up recommendations for children with Kawasaki disease

Risk level	Pharmacological therapy	Physical activity	Follow-up and diagnostic testing	Invasive testing
Level I (no coronary artery changes at any stage of illness)	None beyond first 6–8 weeks	No restrictions beyond first 6–8 weeks	Cardiovascular risk assessment, counselling at 5-year intervals	None recommended
Level II (transient coronary artery ectasia that disappears within 6–8 weeks)	None beyond first 6–8 weeks	No restrictions beyond first 6–8 weeks	Cardiovascular risk assessment, counselling at 3-year to 5–year intervals	None recommended
Level III (one small-medium coronary artery aneurysm/major coronary artery)	Low-dose aspirin (3–5 mg/kg aspirin per day), at least until aneurysm regression documented	For patients <11y old, no restriction beyond 1st 6–8 weeks; patients 11– 20 years old, physical activity guided by biennial stress test, myocardial perfusion scan; contact or high-impact sports	Annual cardiology follow-up with echocardiogram +ECG, combined with cardiovascular risk assessment, counselling; biennial stress test/evaluation of myocardial perfusion scan;	Angiography, if non-invasive test suggests ischaemia

Risk level	Pharmacological therapy	Physical activity	Follow-up and diagnostic testing	Invasive testing
		discouraged for patients taking antiplatelet agents	consider CAA imaging using CT or MR angiography	
Level IV (>1 large or giant coronary artery aneurysm, or multiple or complex aneurysms in same coronary artery, without obstruction)	Long-term antiplatelet therapy combined with warfarin (target INR 2.0–2.5) or low molecular-weight heparin (target: antifactor Xa level 0.5–1.0 U/mL) should be considered in all patients with giant aneurysms	Contact or high-impact sports should be avoided because of risk of bleeding; other physical activity recommendations guided by stress test/evaluation of myocardial perfusion scan outcome	Biannual follow-up with echocardiogram +ECG; annual stress test/evaluation of myocardial perfusion scan 1st angiography at 6–12 mo or sooner if clinically indicated; repeated angiography if non-invasive test, clinical, or laboratory findings suggest ischemia; elective repeat angiography under some circumstances; consider CAA imaging using CT or MR angiography	1st angiography at 6–12 months or sooner if clinically indicated; repeated angiography if non-invasive test, clinical, or laboratory findings suggest ischaemia; elective repeat angiography under some circumstances
Level V (coronary artery obstruction)	Long-term low-dose aspirin; warfarin or low molecular-weight heparin if giant aneurysm persists; consider TPA to dissolve clot; consider use of β-blockers to reduce myocardial O2 consumption; consider statins and/or ACE inhibitors	Contact or high-impact sports should be avoided because of risk of bleeding; other physical activity recommendations guided by stress test/myocardial perfusion scan outcome	Biannual follow-up with echocardiogram and ECG; annual stress test/evaluation of myocardial perfusion scan	Angiography recommended to address therapeutic options; consider CAA imaging using CT or MR angiography intermittently to monitor

CAA, coronary artery aneurysms;; TPA, tissue plasminogen activator.

References:
Lancet 2012:379:1613-1620; *Arch Dis Child* 2014;99:74-83;
http://heart.bmj.com/content/early/2012/10/18/heartjnl-2012-302407.full.pdf+html
http://circ.ahajournals.org/content/110/17/2747.full

What dose of aspirin should be used in the initial treatment of Kawasaki disease? Arch Dis Child 2017;**102**:1180–1182. doi:10.1136/archdischild-2017-313538.
Diagnosis, Treatment and Long-Term Management of Kawasaki Disease. *Circulation.* 2017;135:e927–e999. DOI: 10.1161/CIR.0000000000000484

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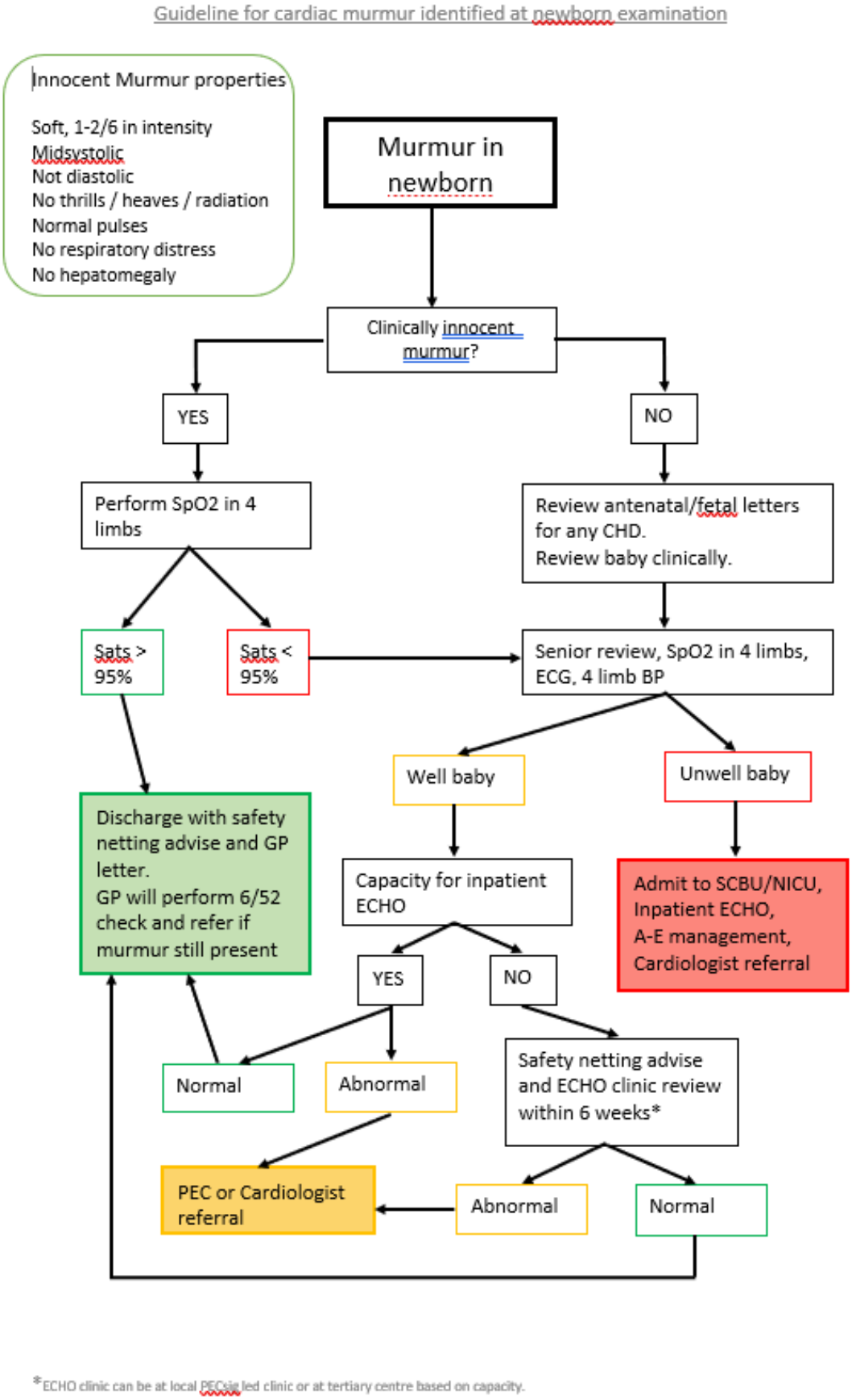
Next review: 2026

6.17 Murmur in a newborn baby



6.18 MRSA and MSSA Colonisation and Infection

All patients being transferred to Bristol for surgery should have throat and nasal swabs for MRSA and MSSA. The request form must specify “Bristol pre-op screening protocol” – it is advisable to telephone the lab to ensure the samples are run as requested.

Methicillin resistant *Staphylococcus aureus* (MRSA) is resistant to all beta-lactam antibiotics (penicillins, cephalosporins) and may be resistant to other classes of antibiotics (multiple-resistant MRSA). Some strains of MRSA are epidemic in character and may cause serious outbreaks of infection in hospitals. MRSA can colonise patients, staff and the hospital environment. Once established in a hospital, MRSA may never be eradicated. The single most important measure to prevent and contain MRSA is meticulous hand hygiene, **particularly with alcohol hand rubs**. Each situation must be dealt with individually and more detailed advice should be obtained from the Infection Control Team.

Patients with MRSA colonisation or infection prior to cardiac surgery **must be highlighted to the surgical team**. Precautionary measures such as eradication measures, final slots on the operating list and isolation cubicles may be necessary.

MSSA colonisation of the nasopharynx is associated with an increased risk of invasive post-operative staphylococcal infection.
If MRSA or MSSA is identified pre-operatively the following measures are taken:

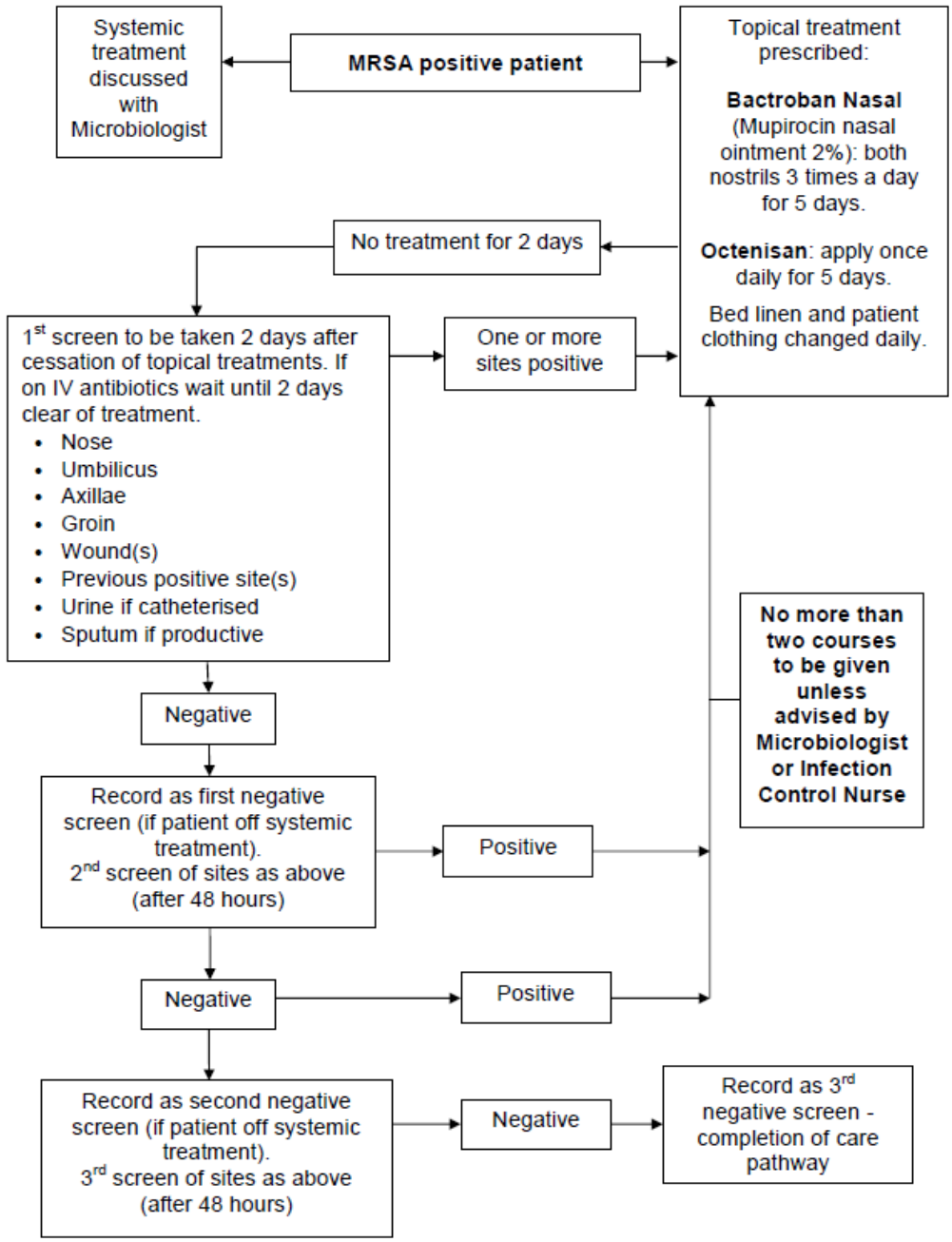
- Mupirocin 2% nasal ointment (Bactroban®) to inner surface of both anterior nares three times a day
- Octenidine dihydrochloride 0.3% (Octenisan®) to skin daily (or any anti-microbial skin wash)
- Continue for 5 consecutive days, (day 5 should be the day of procedure / operation for MSSA patients)

After the 5 days treatment, wait 48 hours then re-swab. If the patient is on IV antibiotics wait until 48 hours after the IV antibiotics have finished before re-swabbing (MSSA patients do not require re-swabbing prior to surgery if they have been treated as described above).
Bristol has an MRSA guideline that should be followed and filled in.

Refer to flow chart below and see Z:\PaedCard\shared\Liaison Nurse Info\MRSA-BRISTOL-GUIDELINES

University Hospitals Bristol NHS
NHS Foundation Trust

Flow Chart for Paediatric/Neonatal Treatment and Screening



If discharged/transferred at any point during treatment please follow guidance detailed further in -

Contributors: Claire Logan, Dirk Wilson – due for review 2026

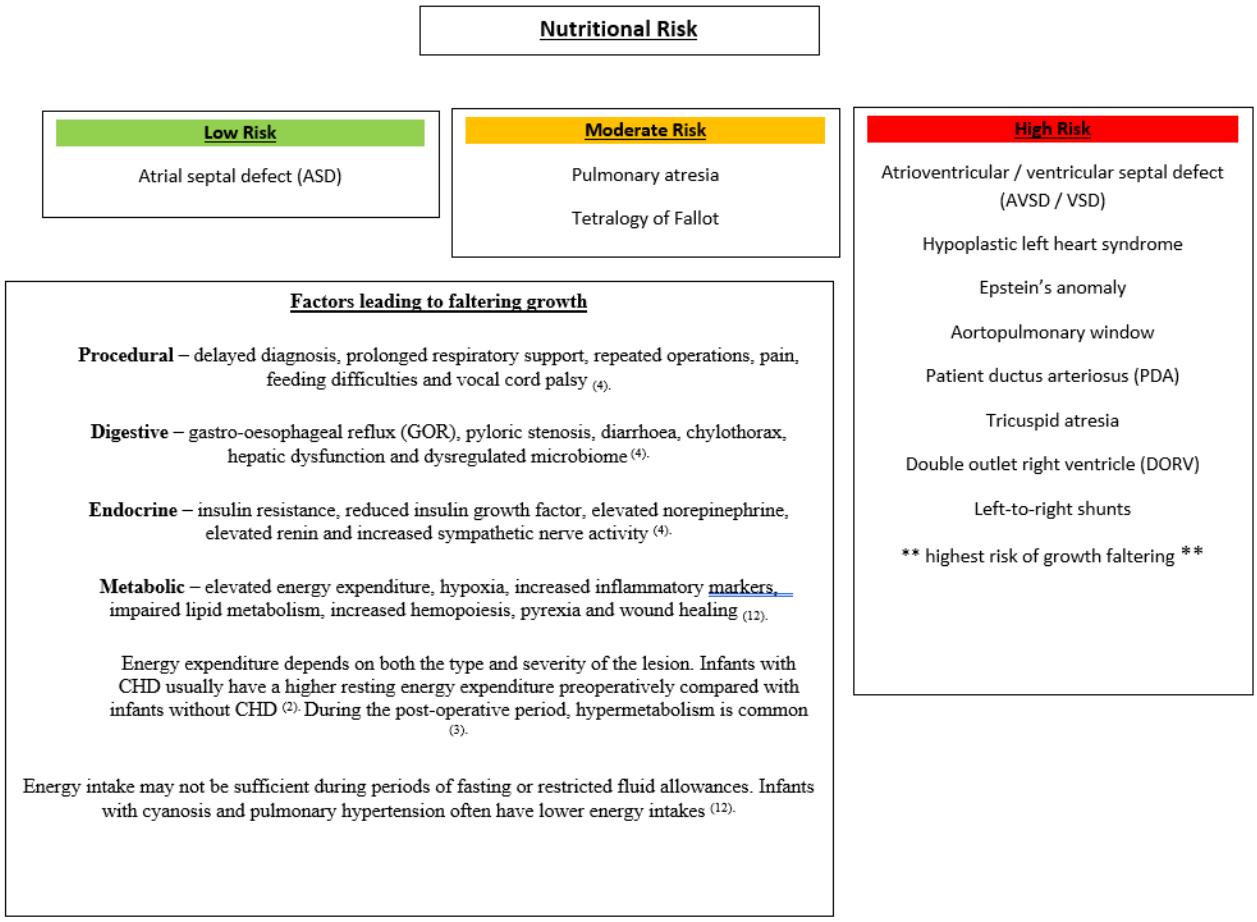
6.19 Nutrition in Cardiac Patients

Aim

The aim of this guideline is to provide clear guidance on optimal feeding strategies for infants and children diagnosed with congenital heart disease (CHD). The goal is to improve growth, development and overall health outcomes.

Background

Children with CHD are at risk of malnutrition and faltering growth.[1] There are many barriers to their ability to consume, absorb and metabolise feeds such as oro-pharyngeal dysfunction, feed intolerance, fluid restriction and impaired absorption.[3] Malnutrition reduces ventricular mass and cardiac output, which may exacerbate an already compromised circulation[4]. Those who are underweight at surgery or fail to gain weight post surgically have an increased risk of mortality.[5]



6.19.1 Growth monitoring

Regular anthropometric measuring is essential in cardiac patients.

On the ward

- Babies are weighed twice weekly (often on request).
- Length is usually done on request - ideally every 2-4 weeks to ensure appropriate growth. Note: lesions that cause cyanosis tend to be associated with stunting (4).

- All babies with congenital health disease and poor growth must be referred to the dietitian.

In the community

- Health visitors weigh children frequently, often more if requested by the dietitian.
- Those with a high risk of developing malnutrition will be on a home monitoring programme. This means that they get weighed twice weekly at home. The cardiologists will decide who needs close monitoring and parents will be issued with weighing scales for discharge.

Calorie Requirement

- If no faltering growth is present and the baby has a low nutritional risk cardiac condition, they may only require 96-120kcal / kg / day [(11)].
- If weight is faltering, a reasonable starting point could be around 120-130kcal/kg/day
- In severe cases of faltering growth, some may require 150+ kcal/kg/day.[(11)]

Micronutrient monitoring

- Chronic electrolyte depletion may become a growth limiting factor.
- It is important to consider whether nutrients such as sodium, potassium, zinc, magnesium, phosphorus or iron may be insufficient in the diet to support adequate growth.[(6)]
- Diuretic control may need to be reviewed. Loop diuretics are commonly used in PICU following surgery to prevent fluid overload, but by nature increase sodium and potassium excretion [(4)].

Feeding Strategies

Breastfeeding:

- Breastfeeding is the initial feed of choice for infants with CHD and should be promoted where possible.
- Breastfeeding may reduce the risk of NEC.[(7)]
- Direct breastfeeding has been found to cause less cardiorespiratory stress than bottle feeding in infants with CHD.[(7)]
- If there are growth concerns, refer to the dietitian for consideration of alternative strategies including use of hind milk or high energy formula.

Promoting maternal milk supply:

- Provide education around the importance of breastfeeding e.g. reduced risk NEC / reduced infections.
- Provide the mother with necessary equipment to support milk expression and storage.
- Ensure mother is eating 3 x meals per day, snacks and is well hydrated. Mothers who breastfeed are entitled to hospital meals on the ward.
- For breastfeeding support and advice, visit: Breastfeeding in the UK - Baby Friendly Initiative (https://www.unicef.org.uk/).

Maximising Calorie Intake:

- A high energy infant formula should be started if weight is faltering or if fluid allowance is restricted.
- The concentration of a standard formula could be increased (specialist dietetic advice is required – not all formulas are suitable).
- Extra calories in the form of fat emulsions / glucose polymers could be added to a standard formula. These supplements should be increased slowly, usually from 2-6%.

There is a fine balance between giving feeds to a sufficient energy density to achieve adequate growth, whilst limiting vomiting and malabsorption

Please liaise with the dietitian regarding high calorie feeds and supplementation.

High calorie infant formulas include:

- Similac High Energy: 100kcal and 2.6g protein / 100ml
- Infatrini Peptisorb: 100kcal and 2.6g protein / 100ml
- SMA High Energy: 100kcal and 2.6g protein / 100ml

Hydrolysed Protein Formula:

- A wide variety is available and specialist dietetic advice is required.
- Extensively hydrolysed formulas are useful in those with cow's milk protein allergy
- May be useful in gastro-oesophageal reflux.
- High energy extensively hydrolysed whey protein (peptide based) formula's such as Infatrini Peptisorb could be started.

Monogen:

- Monogen is a whey protein- based infant formula with low LCT content and high MCT content that may be used in infants with chylothorax.
- The fat sources are from coconut and walnut oil.
- Please speak with the dietitian regarding the initiation of Monogen.

Enteral Tube Feeding:

- Cardiac infants may require NGT feeding at some stage to help them meet growth targets.
- Feeds are often given as 3 hourly bolus feeds, and then responsive feeding can be established once the infant is on their full feed volume.
- If tolerance is a problem / the infant is struggling with increased work of breathing, smaller volumes of feed can be given either 2 hourly or continuously via a pump.
- Jejunal feeding may be indicated in those with gastroparesis, reflux, faltering growth or those at risk of aspiration.[12]
- Be aware that parents must be pump trained if their child receives pump feeding. The dietitian can organise ward based training and will order feeding equipment, ancillaries and feed to their home address.

Gastrostomy Tube Feeding:

- This should be considered in infants requiring long term tube feeding.
- Gastrostomy feeding may be less of an impediment to the development of oral feeding skills than NG feeding.
- Laparoscopic gastro-jejunal feeding tubes have been successfully placed in infants with severe cardiac anomalies and GOR disease and may prevent vomiting [10].

Feeding Consideration:

- The ongoing need for enteral feeding and readiness for oral feeding should be continuously assessed.
- It is recommended to encourage some oral feeding if appropriate to help babies develop the suck / swallow mechanisms.
- Some babies may be capped to only have a small amount of bottled milk, as excessive drinking may increase worsening of breath, which may increase energy expenditure.
- There is evidence that infants receiving enteral nutrition following corrective heart surgery are at increased risk of neurodevelopmental delay and swallowing problems.[8]
- Vocal cord dysfunction strongly correlates with feeding difficulties in children who undergo surgery for CHD.[9]
- A Speech and Language Therapist may need to evaluate the child's eating ability and capability. They may offer dummy dips to promote oral feeding skills.
- High calorie food fortification weaning advice may be provided to prevent weight loss post tube feeding.

6.19.2 Gastro-Oesophageal Reflux (GOR)

- A common problem in cardiac babies.
- GOR disease may lead to anaemia, faltering growth, food refusal, vomiting, pain and haematemesis due to oesophagitis.
- If GOR develops, positioning and feed thickeners may be used as a first-line treatment.

Dietary considerations:

- If bottle / NGT fed – reduce feed volumes if these are excessive for the baby's weight.
- Trial smaller, more frequent feeds.
- Offer a trial of thickened formula (e.g. with added rice starch, corn-starch, locust bean gum or carob bean gum).
- If the above is unsuccessful, consider alginate therapy for a 1-2-week trial.

More severe reflux (GORD) may require:

- H2 antagonist
- Proton pump inhibitors (PPI's)

Although H2 antagonists and PPI's may not prevent GORD, the reduction in acid enables the infant to feel more comfortable and more likely to continue oral feeding.

Enteral tube feeding for GORD:

- Consider enteral tube feeding to promote weight gain in infants with overt regurgitation and faltering growth.
- Jejunal feeding may be considered as an alternative feeding method.

See NICE guidance: https://www.nice.org.uk/guidance/ng1

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Contributors: Sophie Khen and Caroline Powell – due for review 2026

6.20 Oxygen Therapy in Cardiac Patients

Indications for Home Oxygen Therapy include:

- Pulmonary arterial hypertension
- Concurrent lung disease

When it is decided that home O_2 therapy is needed, a referral should be made to the paediatric respiratory team.

- If there is a suspicion of concurrent lung disease, the referral should be made to the consultant.
- Where the indication for home O_2 therapy is primarily cardiac, the referral should be made to the paediatric respiratory nurses. An oxygen prescription form should accompany the referral and this should specify:
    - The desired O_2 flow rate (usually up to 2 L/min if the indication is PHT)
    - The frequency and type of monitoring (e.g. no monitoring, spot checks, overnight pulse oximetry, combination of spot checks and overnight monitoring)
- When changes to home O_2 therapy are desired (e.g. O_2 flow rate, type of monitoring), the paediatric respiratory nurses should be informed and a new prescription form should be issued.

6.21 Pacing in Children and Adolescents

See also section 3.1.3 – Pacing Clinic and 4.4 Temporary Pacing

Indications for pacing in children and adolescents are summarized below – see: https://www.heartrhythmjournal.com/article/S1547-5271(21)01930-5/fulltext

1. Isolated sinus node dysfunction:

	Recommendations	
COR	Isolated Sinus Node Dysfunction	LOE
I	Permanent atrial or dual-chamber pacemaker implantation is indicated for SND when there is correlation of symptoms with age-inappropriate bradycardia.	B-NR
I	Permanent pacemaker implantation is indicated in patients with symptomatic SND secondary to chronic medical therapy for which there is no alternative treatment.	C-EO
IIa	Permanent pacemaker implantation (with rate-responsive programming) is reasonable in patients with symptoms temporally associated with observed chronotropic incompetence.	C-LD
IIb	Permanent pacemaker implantation may be considered in patients with SND and symptoms that are likely attributable to bradycardia or prolonged pauses without conclusive evidence correlating the symptoms with bradycardia following a thorough investigation.	C-EO
III No Benefit	Permanent pacemaker implantation is not indicated in patients with asymptomatic SND.	C-EO
III Harm	Permanent pacemaker implantation is not indicated in patients with symptomatic SND due to a reversible cause.	C-EO

2. Isolated congenital complete AV block:

COR	Recommendations Isolated Congenital Complete Atrioventricular Block	LOE
I	Permanent pacemaker implantation is indicated for patients with CCAVB with symptomatic bradycardia.	B-NR
I	Permanent pacemaker implantation is indicated for patients with CCAVB with a wide QRS escape rhythm, complex ventricular ectopy, or ventricular dysfunction.	B-NR
I	Permanent pacemaker implantation is indicated for CCAVB in asymptomatic neonates or infants when the mean ventricular rate is ≤50 bpm. Ventricular rate alone should not be used as implant criteria, as symptoms due to low cardiac output may occur at faster heart rates.	C-LD
IIa	Permanent pacemaker implantation is reasonable for asymptomatic CCAVB beyond the first year of life when the mean ventricular rate is <50 bpm or there are prolonged pauses in ventricular rate.	B-NR
IIa	Permanent pacemaker implantation is reasonable for CCAVB with left ventricular dilation (z score ≥3) associated with significant mitral insufficiency or systolic dysfunction.	C-LD
IIb	Permanent pacemaker implantation may be considered for CCAVB in asymptomatic adolescents with an acceptable ventricular rate, a narrow QRS complex, and normal ventricular function, based on an individualized consideration of the risk/benefit ratio.	C-LD

3. AV block – other considerations:

COR	Recommendations Atrioventricular Block: Other Considerations	LOE
I	Permanent pacemaker implantation is indicated in patients with clinically significant ventricular tachycardia (VT) that is pause dependent or associated with severe bradycardia; ICD implantation may be considered as a reasonable alternative.	C-LD
I	Permanent pacing is indicated in *symptomatic* patients with idiopathic advanced second- or third-degree AV block not attributable to reversible causes.	C-LD
IIa	Permanent pacemaker implantation is reasonable for any degree of AV block that progresses to advanced second- or third-degree with exercise in the absence of reversible causes.	C-LD
IIb	Permanent pacemaker implantation may be considered for patients with intermittent advanced second- or third-degree AV block not attributable to reversible causes and associated with minimal symptoms that are otherwise unexplained.	C-LD
III Harm	Permanent pacemaker implantation is not indicated for asymptomatic first-degree AV block or asymptomatic second-degree Mobitz type I.	C-LD

4. Post-operative AV block:

COR	Recommendations Postoperative Atrioventricular Block	LOE
I	Permanent pacemaker implantation is indicated for postoperative advanced second- or third-degree AV block that persists for at least 7–10 days after cardiac surgery.	B-NR
I	Permanent pacemaker implantation is indicated for late-onset advanced second- or third-degree AV block especially when there is a prior history of transient postoperative AV block.	C-LD
IIb	Permanent pacemaker implantation may be considered for unexplained syncope in patients with a history of transient postoperative advanced second- or third-degree AV block.	C-LD
IIb	Permanent pacemaker implantation may be considered at <7 postoperative days when advanced second- or third-degree AV block is not expected to resolve due to extensive injury to the cardiac conduction system.	C-EO
IIb	Permanent pacemaker implantation may be considered in select patients with transient postoperative advanced second- or third-degree AV block who are predisposed to progressive conduction abnormalities (see text).	C-EO

5. Congenital heart disease – specific considerations:

COR	Recommendations Congenital Heart Disease *All the recommendations in children with a structurally normal heart apply, but in addition:*	LOE
I	Permanent pacemaker implantation is indicated for CCAVB in neonates or infants with complex CHD when bradycardia is associated with hemodynamic compromise or when the mean ventricular rate is <60–70 bpm.	C-LD
IIa	Permanent pacemaker implantation with atrial antitachycardia pacing is reasonable for patients with CHD and recurrent episodes of intra-atrial re-entrant tachycardia when catheter ablation or medication are ineffective or not acceptable treatments.	B-NR
IIa	Permanent atrial or dual-chamber pacemaker implantation is reasonable for patients with CHD and impaired hemodynamics due to sinus bradycardia or loss of AV synchrony.	C-LD
IIa	Permanent atrial or dual-chamber pacing is reasonable for patients with tachy-brady syndrome and symptoms attributable to pauses due to sudden-onset bradycardia.	C-LD
IIa	Permanent pacemaker implantation is reasonable for sinus or junctional bradycardia with *complex* CHD when the mean awake resting heart rate is <40 bpm or when there are prolonged pauses in the ventricular rate.	C-EO
IIb	Permanent pacing may be considered for sinus or junctional bradycardia with *simple or moderate* CHD when the mean awake resting heart rate is <40 bpm or when there are prolonged pauses in the ventricular rate.	C-EO
III Harm	Endocardial leads should be avoided in patients with CHD and intracardiac shunt except in select cases, for whom there should be an individualized consideration of the risk/benefit ratio. In these exceptional cases anticoagulation is mandatory, but thromboembolism remains a risk.	B-NR

6. Neuromuscular disease and other progressive condution system disease:

COR	Recommendations Neuromuscular Diseases and Other Progressive Cardiac Conduction Diseases	LOE
I	Permanent pacemaker implantation is indicated in patients with neuromuscular diseases with symptomatic bradycardia due to SND or any degree of AV block.	B-NR
I	Permanent pacemaker implantation is indicated in Kearns-Sayre syndrome for any degree of AV block (including first-degree AV block) and/or conduction abnormality because of unpredictable progression of conduction disease.	C-LD
IIa	Permanent pacemaker implantation is reasonable in patients with myotonic dystrophy type 1 for marked first-degree AV block (PR interval >240 ms) or intraventricular conduction delay (native QRS duration >120 ms). Additional defibrillator capability may be considered.	B-NR
IIa	Permanent pacemaker implantation is reasonable in patients with lamin A/C gene mutations, including limb-girdle and Emery-Dreifuss muscular dystrophies with a PR interval >240 ms and/or left bundle branch block. Additional defibrillator capability may be considered.	C-LD
IIb	Permanent pacemaker implantation may be considered for any patient with any progressive cardiac conduction disease with potential for rapid deterioration of AV nodal function, even in the presence of normal AV conduction after taking into consideration patient age, size, and other individual risk factors.	C-LD

7. Vasovagal (neurocardiogenic syncope):

COR	Recommendations Neurocardiogenic Syncope	LOE
IIa	Permanent pacemaker implantation is reasonable with severe recurrent breath-holding spells with documentation of cardioinhibitory response on ECG monitoring and complicated by prolonged syncope, prolonged postanoxic convulsions, and other bradycardia-induced symptoms.	B-NR
IIb	Permanent pacing may be considered for recurrent symptomatic neurocardiogenic syncope associated with documented spontaneous bradycardia or asystole in patients who have failed other medical treatments.	C-LD
IIb	Permanent pacemaker implantation may be considered in patients with epilepsy associated with severe symptomatic bradycardia (ictal induced) who have failed to improve with antiepileptic medical therapy.	C-LD
III No benefit	Permanent pacing is not indicated for neurocardiogenic syncope solely on the basis of a positive cardioinhibitory tilt response.	C-EO
III Harm	Permanent pacing is not indicated for neurocardiogenic syncope with hypotension as the major or significant component of the symptoms.	C-EO

8. Cardiac channelopathies:

COR	Recommendations Cardiac Channelopathies	LOE
I	Permanent pacemaker implantation is indicated in channelopathy patients with pause-dependent, clinically significant VT; ICD implantation may be considered as a reasonable alternative.	C-LD
IIb	Permanent pacemaker implantation may be considered as adjunctive therapy in patients with long QT syndrome and functional 2:1 AV block.	C-LD
IIb	Permanent pacemaker implantation may be considered as adjunctive therapy in patients with long QT syndrome or other channelopathies where a faster heart rate may decrease the arrhythmia burden or symptoms due to bradycardia.	C-LD
III No benefit	Atrial pacing alone is not indicated in patients with complete atrial standstill due to the high potential for noncapture of the myocardium.	C-LD

9. Inflammation/Infection:

COR	Recommendations Inflammation/Infection	LOE
I	Permanent pacing is indicated in patients with high-grade or symptomatic AV block attributable to a known potentially reversible cause when AV block does not resolve despite treatment of the underlying cause.	C-LD
IIa	Pacemaker implantation is reasonable in Chagas disease and advanced second- or third-degree AV block, as spontaneous resolution is unlikely. ICD implantation may be a reasonable alternative.	C-LD
III No benefit	Permanent pacing should not be performed in patients who had acute AV block attributable to a known reversible cause, when there is recovery of normal AV conduction.	C-EO

See PACES guideline for ICD indications in children:
[2021 PACES Expert Consensus Statement on the Indications and Management of Cardiovascular Implantable Electronic Devices in Pediatric Patients: Executive Summary - Heart Rhythm (heartrhythmjournal.com)]

Pacing Terminology

Position I	Position II	Position III	Position IV	Position V
Paced chamber	**Sensed chamber**	**Mode of response**	**Programmability**	**Anti-tachycardia capability**
O – none	O – none	O – none	O – none	O – none
A – atrium	A – atrium	T – triggered	M – multiprogrammable	P – pacing
V – ventricle	V – ventricle	I – inhibited	C - communicating	S – shock
D – dual (A+V)	D – dual (A+V)	D – dual (T+I)	R – rate modulation	D – dual (P+S)

Common pacing modes in children are:
VVI Paces ventricle
Senses intrinsic ventricular activity
Inhibited by intrinsic ventricular activity

VVIR Paces ventricle

Senses intrinsic ventricular activity
Inhibited by intrinsic ventricular activity
Rate response mode is on (heart rate will increase with physical activity)

DDD Paces ventricle and atrium
Senses intrinsic atrial and ventricular activity
Triggered and Inhibited by intrinsic atrial ventricular activity

NB – patient's physical activity will lead to an increase in SA node discharge, so heart rate will increase.

6.22 Persistent Ductus Arteriosus in Premature Neonates

PDA is a common finding in premature infants and their day-to-day clinical management rests with the neonatal team. Neonates referred with suspected PDA should be managed as any other inpatient referral (see Section 2.2.2, and the full guidance on the SWSW-CHD Network website:
https://www.swswchd.co.uk/image/page/04%20Paed%20%20%20PDA%20%20GuidanceForReferralForCardiologicalAssessmentAndPo-1.pdf

In some cases the neonatal team will request referral to the congenital heart disease surgical team for PDA ligation. In these cases, the Wales/Southwest pathway for PDA ligation needs to be followed (see below).

PDA Ligation in preterm infants carries a significant risk of mortality (up to 10%) as well as significant risk of post-operative morbidity. Therefore it is a procedure that should only be considered in infants for whom the alternative (i.e. not ligating the ductus) is deemed by the treating doctors to carry a higher risk of adverse outcome.
The published data on outcomes for infants in whom ligation has or has not been performed does not permit a simple evidence-based approach to identify those infants for whom ligation is most appropriate. Therefore, ligation of a PDA should only be considered for the following groups of infants, and (except in unusual circumstances) when the following conditions have been met:

- The infant has a "*clinically significant*" PDA (see below)
- The infant was born at 30 weeks gestation or less
- The infant is now 3 weeks or more of age
- The infant is currently ventilator-dependent and not making clear progress towards extubation**
- Treatment with indometacin/ibuprofen has been considered and at least one course (usually two) has been given if appropriate unless contra-indications to treatment present.
- The infant has already received a course of steroid treatment for chronic lung disease, or the treating neonatal team consider that there is a contra-indication to this treatment
- The infant has been given at least a short course of diuretic therapy to improve pulmonary compliance
- There is currently no evidence of untreated infection or NEC

**Although the great majority of infants referred for PDA ligation will be ventilator dependent, a small group may be considered for ligation if they are on high level FiO_2 and/or CPAP and cannot be weaned, and/or they are not tolerating sufficient enteral nutrition to grow and the PDA is felt to be the main factor underlying this problem.

Almost all infants who are to be considered for PDA ligation should meet the criteria, or have an identifiable contra-indication to such treatment options.

See Section 5.10.4 for echo assessment of PDA. **Contributors: Jon Forsey, Orhan Uzun, Mark Walsh, Dirk Wilson**

6.23 Protein-losing enteropathy (PLE) / Plastic bronchitis (PB) after Fontan

Definition/Incidence/Aetiology

PLE is a condition characterised by excessive gastrointestinal protein loss. The liver is unable to compensate and this leads to hypoalbuminaemia, hyopoproteinaemia and reduced serum immunoglobulins. Cardiac causes include the Fontan circulation (10-15% of patient long-term), severe CCF and constrictive pericarditis. The risk factors for PLE following the Fontan operation include:

- Systemic RV
- Perioperative renal failure
- High venous pressures
- History of perioperative chylothorax or chyloperitoneum
- Preoperative infection
- ?Lack of fenestration

Clinical features

- Oedema (facial and lower body)
- Abdominal distension
- Ascites
- Anorexia
- Loose stools/flatus

Any report of these symptoms in Fontan patients should prompt investigation.

Plastic Bronchitis is the pulmonary equivalent of PLE. The features are:

- Cough and choking
- The expectoration of tenacious material which, if spread out, takes the shape of the bronchial tree – this is a "cast" – the material is proteinaceous

Laboratory and Other Investigations

It may be advisable to perform annual LFTs in all postoperative Fontan patients.
Patients with clinical features should have the following investigations:

- FBC (with differential)
- U&E/creat/LFT/Ca^{2+}
- Serum immunoglobulins
- Stool vs serum α-1 antitrypsin
- Urinary protein (to rule out renal protein loss)
- ECG/24 hour tape – to rule out arrhythmia
- Echo – to rule out haemodynamic cause
- Cardiac catheterisation – to rule out haemodynamic cause and consider intervention (e.g. transcatheter fenestration)
- ?MRI (relationship of lateral tunnel to native RA)
- Consider need for GI investigations, including endoscopy, to exclude primary GI causes
- Baseline bone densitometry studies (dexa scan) – heparin and prednisolone, which are both used to treat PLE, cause bone demineralisation.

Treatment

PLE and PB are complex clinical problems and are managed on a case-by-case basis with close involvement of the surgical centre ($\pm$ other tertiary centre such as GOS or Birmingham).
General supportive measures include:

- High-protein, MCT diet

- ACE inhibitors + diuretics should be considered, particularly if there is significant AV valve regurgitation
- Consideration of sildenafil (to reduce PVR)

Anti-PLE treatments also include:

- High-dose steroids (2 mg/kg per day, tail according to response)
- Oral budesonide (Budenofalk capsules, 3 mg by mouth three times daily for up to 8 weeks – review effect and consider reducing dose gradually over 2 weeks)
- Subcutaneous unfractionated (standard) heparin injections (the dose is usually sub-therapeutic [1500 unit subcutaneous bd, regardless of weight] and warfarin should be continued – monitor KCCT and INR (NB – follow up bone density needed)
- Consider IV albumin if the serum albumin is < 25 and peripheral oedema is severe
- Septrin if evidence of immunosupression
- Calcium supplementation
- Aggressive management of infection (increased risk due to immunosuppression).

Anti-PB treatments also include:

- Nebulised steroids
- Nebulised normal saline, TPA, urokinase, or N-acetyl cysteine
- Chest physiotherapy
- Bronchoscopy

Therapeutic interventions for PLE and PB include:

- Addressing any haemodynamic problem } transcatheter or
- Fenestration } operative
- Transplantation (but in up to 50% of cases the PLE persists)

Outcomes

The 5 year mortality in patients with a venous pressure of 16 mmHg is 50%.
25% of patients may respond to general supportive measures and anti-PLE medications.
Papers suggest that surgical outcomes are poor, but this may relate to case selection.
Patients with PLE, therefore, require close surveillance.

Contributors: Orhan Uzun, Dirk Wilson – reviewed 2023

6.24 Pulmonary Hypertension in Childhood

6.24.1 Definition, Classification and WHO Functional Status

See
https://academic.oup.com/eurheartj/article/43/38/3618/6673929?login=false
(click on the link on the website for the PDF version)

Pulmonary Hypertension
The normal range for pulmonary artery (PA) pressure is dependent on age. In general, resting peak PA pressures should be around 1/3 of systemic blood pressure (i.e. systolic [ressure of 25-40 mmHg depending on age) and mean PA pressure should be no more than 20 mmHg). Pressures in excess of these values indicate pulmonary hypertension.

Haemodynamic definitions of PAH

Definition	Haemodynamic characteristics
PH	mPAP >20 mmHg
Pre-capillary PH	mPAP >20 mmHg PAWP ≤15 mmHg PVR >2 WU
Isolated post-capillary PH	mPAP >20 mmHg PAWP >15 mmHg PVR ≤2 WU
Combined post- and pre-capillary PH	mPAP >20 mmHg PAWP >15 mmHg PVR >2 WU
Exercise PH	mPAP/CO slope between rest and exercise >3 mmHg/L/min

Classification of pulmonary hypertension

GROUP 1 Pulmonary arterial hypertension (PAH)
1.1 Idiopathic 1.1.1 Non-responders at vasoreactivity testing 1.1.2 Acute responders at vasoreactivity testing
1.2 Heritable
1.3 Associated with drugs and toxins
1.4 Associated with: 1.4.1 Connective tissue disease 1.4.2 HIV infection 1.4.3 Portal hypertension 1.4.4 Congenital heart disease 1.4.5 Schistosomiasis
1.5 PAH with features of venous/capillary (PVOD/PCH) involvement
1.6 Persistent PH of the newborn

GROUP 2 PH associated with left heart disease
2.1 Heart failure:
2.1.1 with preserved ejection fraction
2.1.2 with reduced or mildly reduced ejection fraction
2.2 Valvular heart disease
2.3 Congenital/acquired cardiovascular conditions leading to post-capillary PH
GROUP 3 PH associated with lung diseases and/or hypoxia
3.1 Obstructive lung disease or emphysema
3.2 Restrictive lung disease
3.3 Lung disease with mixed restrictive/obstructive pattern
3.4 Hypoventilation syndromes
3.5 Hypoxia without lung disease (e.g. high altitude)
3.6 Developmental lung disorders
GROUP 4 PH associated with pulmonary artery obstructions
4.1 Chronic thrombo-embolic PH
4.2 Other pulmonary artery obstructions
GROUP 5 PH with unclear and/or multi-factorial mechanisms
5.1 Haematological disorders
5.2 Systemic disorders
5.3 Metabolic disorders
5.4 Chronic renal failure with or without haemodialysis
5.5 Pulmonary tumour thrombotic microangiopathy
5.6 Fibrosing mediastinitis

Most paediatric patients will be in classes:

1.1 Idiopathic PAH
1.2 PAH with genetic cause
1.4 PAH in association with CHD (1.4.4)
1.6 PPHN (see section 6.23.4)
2.2 PH with valve disease
2.3 Post-capillary PH (related to L heart disease)
3 Associated with lung disease
4.1 Thrombo-embolic PH

Functional Status

Class	Description
WHO-FC I	Patients with PH but without resulting limitation of physical activity. Ordinary physical activity does not cause undue dyspnoea or fatigue, chest pain, or near syncope
WHO-FC II	Patients with PH resulting in slight limitation of physical activity. They are comfortable at rest. Ordinary physical activity causes undue dyspnoea or fatigue, chest pain, or near syncope
WHO-FC III	Patients with PH resulting in marked limitation of physical activity. They are comfortable at rest. Less than ordinary activity causes undue dyspnoea or fatigue, chest pain, or near syncope.
WHO-FC IV	Patients with PH with an inability to carry out any physical activity without symptoms. These patients manifest signs of right heart failure. Dyspnoea and/or fatigue may even be present at rest. Discomfort is increased by any physical activity

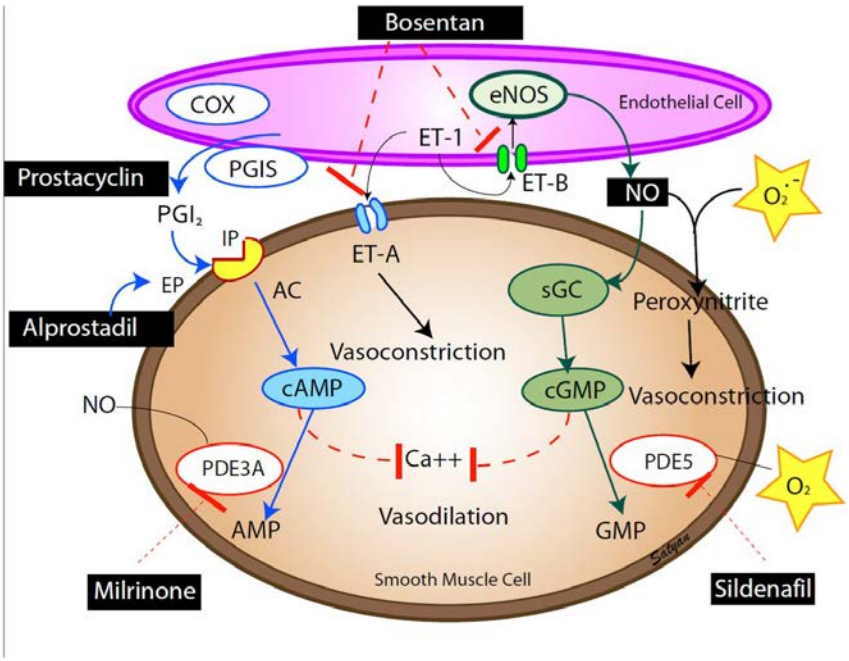
6.24.2 Treatment of Pulmonary Hypertension

Left untreated, the median survival of children with idiopathic or familial pulmonary arterial hypertension is around 4 years. The outlook is better for patients with Eisenmenger syndrome, with many patients surviving beyond their 20s, but developing significant limitation by their 30s. Patients with significant PAH should be referred to the specialist PAH Team from GOS.

Interventional management of Pulmonary Hypertension

Treatment	Action	Evidence Base	Comment
Atrial septostomy	Permits R→L shunting at atrial level, preserving cardiac output at the expense of systemic desaturation	Non-controlled trials have shown improved cardiac output following the procedure	High-risk procedure (mortality up to 16%), only performed in specialist centres.
Potts shunt (descending aorta to LPA connection)	Permits R→L shunting at great vessel level, preserving cardiac output at the expense of systemic desaturation	Non-controlled trials have shown improved cardiac output following the procedure	May be performed surgically or via the transcatheter route. Procedural mortality is 15-30%, so this is reserved for severely affected cases.

Drug treatment of PAH:



https://publications.aap.org/view-large/figure/5960777/neoreviews_12Keszlerfig1.jpeg

Calcium channel blockers

- Amlodipine
- Diltiazem
- Felodipine

Endothelin receptor antagonists

- Ambrisentan
- Bosentan
- Macitentan

Phospodiesterase 5 inhibitors

- Sildenafil
- Tadalafil

Prostacyline analogues

- Oral
    - Beraprost
    - Treprostinil
- Inhaled
    - Iloprost
    - Treprostinil
- IV or SC
    - Epoprostenol
    - Treprostinil

Prostacycline receptor agonist

- Selexipag

Soluble guanylate cyclase stimulator

- Riociguat

Patients may require treatment with more than one form of therapy. These are overseen by the National Pulmonary Hypertension Service, based at Great Ormond Street Hospital.

Contributor: Dirk Wilson

6.24.4 Persistent Pulmonary Hypertension of the Newborn (PPHN)

See NNU guideline:
Neonatology Clinical Portal - PPHN uhw version1.pdf - All Documents (sharepoint.com)

Introduction:
In utero only 10-15% of the cardiac output reaches the lungs via the pulmonary circulation. Oxygenated umbilical venous blood is streamed to the left atrium (and the brain) and the aorta (and the body) by the combined effects of the foramen ovale, the ductus arteriosus and pulmonary arterial vasoconstriction. After delivery, inflation of the lungs and increased PaO_2 are the principle factors promoting pulmonary vasodilatation, reduced pulmonary vascular resistance (PVR) and improved pulmonary perfusion.
Failure to achieve this expected fall in PVR and therefore failure of oxygenation of the venous blood returned to the heart is described by the term Persistent Pulmonary Hypertension of the Newborn (PPHN). PPHN is being increasingly recognised in neonatal practice with an estimated incidence of 2-6/1000 births. It can occur in both term and preterm neonates and is perhaps the most common cause of death in infants of birth weight > 1000g.

Predisposing factors:

- Hypothermia, hypoglycaemia, hypoxia and acidosis
- Bacterial pneumonia, meconium aspiration syndrome, surfactant deficiency lung disease

- Chronic fetal hypoxia, placental insufficiency, postmaturity, polycythaemia
- Congenital diaphragmatic hernia
- Primary pulmonary hypoplasia
- Congenital alveolar capillary dysplasia

Clinical features:
- The most important clinical feature is difficulty in oxygenating the neonate with persistent low O_2 saturations despite increasing FiO_2 and ventilatory support.
  - The blood gas (arterial) is likely to show severe hypoxemia.
  - There is significant difference in pre and post ductal PaO_2 (>2.5 kPa) or O_2 saturations (5-10%).
- A prominent right ventricular impulse may be noted and murmurs due to tricuspid regurgitation or pulmonary regurgitation may be heard.
- Signs of heart failure may be present.

Investigations:
- Refer to NICU guidance
- Echocardiography is the gold standard and should be used to establish the diagnosis and inform management.

Echocardiographic assessment of pulmonary hypertension:

Tricuspid regurgitation:
- RV pressure can be calculated from the TR jet ($4v^2$ + add estimated RA pressure)
- Ensure the envelope is complete – if the patient clinically has severe PPHN and the TR jet is 2-3 m/sec, you are missing something
- Interpret in the context of systemic BP

Atrial shunting and other shunts:
- Some degree of right-to-left atrial shunting through the patent foramen ovale is common, although it is rare for this to be purely right-to-left (Pure right-to-left flow indicates total anomalous pulmonary venous connection [TAPVC] until proved otherwise).
- Bowing of the interatrial septum to the left is commonly seen.
- Right-to-left atrial shunting reflects right atrial filling (diastolic) pressure
- If a VSD is present, bidirectional shunting may be noted.

Ductal flow:
- The direction and velocity of ductal blood flow can give useful information on PAP.
- Pure right-to-left flow indicates PAP is higher than aortic pressure throughout cardiac cycle.
- Bidirectional flow occurs when the aortic and pulmonary pressures are approximately equal. Flow is left-to-right during diastole and right-to-left during systole (as the pulmonary arterial pressure wave reaches the duct before the aortic pressure wave).
- Bidirectional flow is common in healthy babies in the first 12 hours but changes to pure left-to-right when aortic pressures become higher than pulmonary pressures.

Other parameters are reserved for more specialist evaluation and include
- TPV/RVET ratio
- RPEP/RVET ratio
- IVRT (from TV annulus tissue Doppler)
- Pulmonary velocity (or artery) acceleration time – PVAT or PAAT – record pulsed-wave Doppler in the main pulmonary artery. Measure time interval from onset of flow to the peak of flow.

Cardiac function and output:

- There may be enlargement of the RV and RA, as well as the main pulmonary artery.
- There may be flattening (RV:LV pressure >0.5) or even bowing (RV:LV pressure ≥1.0) of the interventricular septum to the left as RV pressure rises.
- As cardiac output is dependent on venous return to the RA and LA, cardiac output (both RVO and LVO) is frequently reduced with PPHN. Severe PPHN may be associated with LVO below 100ml/kg/min (normal 150-300ml/kg/min)
- Quantitative assessment of cardiac function may assist with decisions and assessments of the roles of inotropes and inhaled nitric oxide.
- If the LA and LV appear under-filled, it is critical to exclude TAPVD. Demonstration of a left-to-right shunt at atrial level essentially excludes TAPVD.

Grading of severity of PPHN based on echo parameters

Mild PPHN (PVR high but below SVR)
Clinical parameters in keeping with diagnosis of PPHN.
PFO flow exclusively L→R
PDA flow bidirectional but predominately L→R at low velocity
PAAT ≥ 70 msec

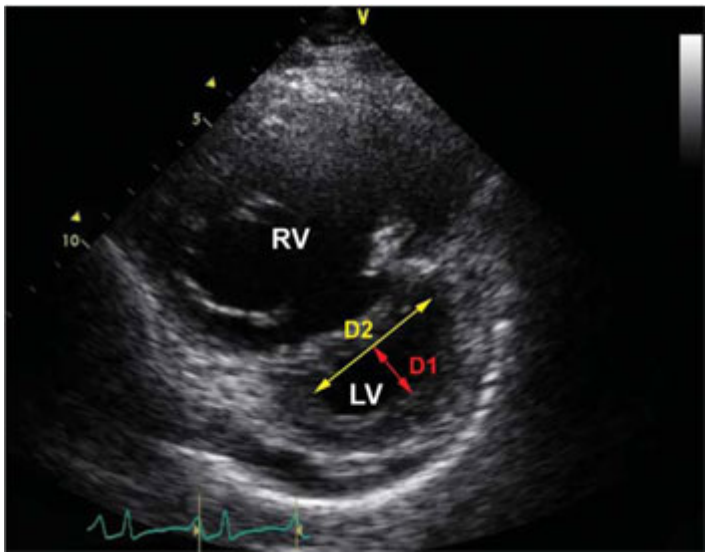
Moderate PPHN (PVR just higher than SVR)
Clinical parameters in keeping with diagnosis of PPHN.
PFO bidirectional, but mainly L→R
PDA flow bidirectional but predominately R→L with peak systolic (R→L) Doppler <1.0 m/sec
PAAT 50-70 msec

Severe PPHN (PVR clearly higher than SVR)
Clinical parameters in keeping with diagnosis of PPHN.
PFO flow bidirectional but predominately R→L
PDA flow exclusively or predominately R→L, with systolic R→L Doppler 1.0 m/sec or greater.
PAAT <50 msec

In all cases the echo report should state:

- Systemic systolic BP
- Estimated systolic PA pressure based on TR jet + estimated RA pressure of 10 or 15 mmHg (depending on IVC collapse and RA size)
- PA acceleration time
- LV eccentricity index (record ratio in systole and diastole to see how useful these are as a rough measure over time)

Guide to LV eccentricity index (a reading of D2/D1 >1.2 in systole is abnormal).:
http://www.phaonlineuniv.org/Journal/Article.cfm?ItemNumber=671



Management of PPHN

Refer to local NNU guidelines.

Milrinone for PPHN

Milrinone works via PDE inhibition and leads to relaxation of vascular smooth muscle. It potentiates the effect of NO. It may improve cardiac diastolic function. Ensure the patient is well filled before instituting treatment.

Dose: 50-75 microgram/kg over 30-60 minutes, followed by infusion of 30-45 microgram/kg/hour for maintenance. Doses of up to 75 microgram/kg/hour may be used. The dose should be reduced in patients with renal failure.

Pulmonary Vasodilators

Inhaled nitric oxide (iNO)

iNO is the first pulmonary vasodilator of choice. Cochrane review has shown that use of iNO in PPHN decreases oxygen index and need for ECMO significantly[6]. It should be started at 20ppm can be added to conventional ventilation or HFOV. Methaemoglobin and Nitrogen dioxide (NO2) levels should be monitored during administration of iNO

Prostacyclin

Prostacyclin acts to elevate cellular cAMP levels. It causes vasodilatation of both pulmonary and systemic circulation and hence systemic hypotension may be a problem. Continuous infusion of 5-20 nanograms/k/min can be used if systemic BP is high enough. Inhaled Prostacyclin (two small studies have shown improvement in oxygenation at doses of 20-50ng/kg/min) may also be used with less systemic hypotensive effect.[7,8]

Sildenafil

It has been shown to be effective in decreasing oxygen index in a randomised controlled trial as well as in observational studies in the treatment of PPHN. [9,10,11] (For dosing see Sildenafil protocol below)

Magnesium

It has been used as a pulmonary vasodilator[12]. Toxicity appears low and the effects on systemic circulation are limited unless used in high dose in the presence of myocardial ischaemia. Aim for plasma levels 2-4mmol/L

Adenosine for PPHN

It acts via adenosine receptors on endothelium, to elevate intracellular cAMP, causing smooth muscle relaxation. Continuous infusion of 25-50mcg/k/min has been used in small studies with success.[13,14]

ECMO

ECMO is used as last resort if above therapies fails to achieve adequate oxygenation (Oxygen Index >40). A policy of using ECMO in mature infants with severe but potentially reversible respiratory failure results in significantly improved survival without increased risk of severe disability amongst survivors. For babies with diaphragmatic hernia ECMO offers short term benefits but the overall effect of employing ECMO in this group is not clear.[15]

Contributor: Orhan Uzun

References:

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3. Clark RH, Yoder BA, Sell MS. Prospective, randomized comparison of high-frequency oscillation and conventional ventilation in candidates for extracorporeal membrane oxygenation. J Pediatr 1994; 124:447-54.
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6. Finer NN, Barrington KJ. Nitric oxide for respiratory failure in infants born at or near term. Cochrane Database Syst Rev. 2006 Oct
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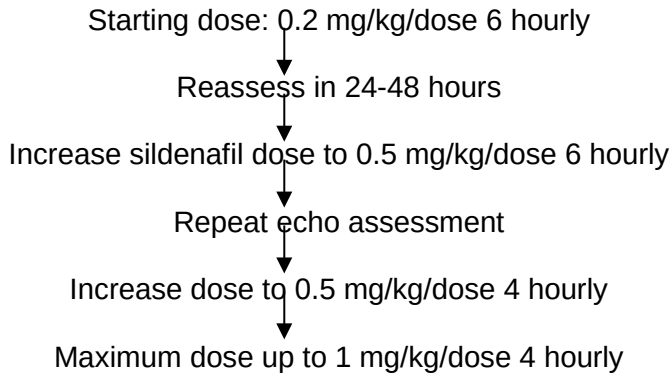
11. Baquero H, Soliz A, Neira F, Venegas ME, Sola A. Oral Sildenafil in infants with persistent pulmonary hypertension of the newborn: a pilot randomized blinded study. Pediatrics. 2007 Jan; 119(1):215-6; author reply 216
12. W D Carroll, R Dhillon Sildenafil as a treatment for pulmonary hypertension Arch. Dis. Child. 2003;88;827-828
13. Ng C, Franklin O, Vaidya M, Pierce C, Petros A. Adenosine infusion for the management of persistent pulmonary hypertension of the newborn. Pediatr Crit Care Med. 2004 Jan; 5(1):10-3.
14. Konduri GG, Garcia DC, Kazzi NJ, Shankaran S. Adenosine infusion improves oxygenation in term infants with respiratory failure. Pediatrics. 1996 Dec; 98(6 Pt 1):1224-5.
15. Elbourne D, Field D, Mugford M. Extracorporeal membrane oxygenation for severe respiratory failure in newborn infants. Cochrane Database of Systematic Reviews 2002
16. PPHN Guideline – Neonatal Unit Clinical Guidelines

Sildenafil Dose Protocol

Sildenafil is being used increasingly, particularly in the newborn period (e.g. PPHN amd chronic lung disease patients). It should be instituted under consultant direction as follows:



NB – **chronic use** of high dose sildenafil in patients with PA hypertension is associated with worse outcomes and the MHRA has issued the following advice:

For patients who weigh ≤ 20 kg the maximum recommended dose is 10 mg three times a day and for patients > 20 kg the maximum recommended dose is 20 mg three times a day.

6.25 PVCs in Children

Premature ventricular complexes (PVCs) are common in children and adults. Often they occur without symptoms, but some patients are aware of PVCs and they may feel unpleasant (“missed beat”, “painful beat”, “bumping sensation”). In most cases they are a benign finding but in some cases they may signify an underlying disease process, such as psot cardiac surgery, cardiomyopathy, e.g. ARVC, or an ion channelopathy, e.g. CPVT. PVCs are commonly seen in patients with mitral valve prolapse. They are more likely to occur with electrolyte disturbance such as hypokalaemia, hypercalcaemia and hypomagnesaemia.

When are PVCs pathological?

More likely to be benign	**More likely to be pathological**
Occur infrequently, <5-10% of total beats on Holter	Occur frequently, >10% of total beats on Holter; more concern if >25%
PVCs have a single, consistent morphology	More than one morphology, or one morphology and occurring >25% of total beats on Holter
PVCs have a LBBB morphology and inferior axis signifying RVOT origin	RBBB morphology (LV origin) or LBBB with non-inferior axis
Ectopics occur as isolated beats	Ectopics occur in couplets, triplets or salvoes, long runs of bigeminy

Investigation of PVCs

If rare ectopic beats are noted in isolation, with LBB morphology, inferior axis, no symptoms and normal heart structure and function on echo, then no further investigation is needed. PVCs with a right bundle-branch block pattern (or S wave with a secondary R wave) in V1 have a left ventricular outflow tract origin and are more likely to be significant.

In patients with the following features, further investigation is warranted:

- Symptoms of palpitation or syncope

- FHx of sudden cardiac death, cardiomyopathy or sustained arrhythmia
- Structural heart disease
- Abnormal ventricular function (RV or LV)

Investigations to consider include:

- Blood tests to rule out electrolyte disturbance
- Holter monitoring (frequency, morphology variation, evidence of VT?)
- Exercise testing (does ectopy disappear or worsen with exercise?)
- MRI scanning (if abnormal cardiac function or texture noted on echo)
- Signal averages ECG (if abnormal, this may be a marker for future development of non-sustained VT)
- Invasive electrophysiological testing

Management of PVCs

In most cases the patient can be reassured that the PVCs are benign and that no further action is needed.

Patients with PVC rates of >10% of total beats should have ongoing clinical review to assess cardiac function (e.g. annually or biannually).

Patients with PVC rates of >10-25% may benefit from therapeutic intervention, particularly if ventricular function is compromised. Treatments to consider include:

- Beta-blocker (non-athletic individual)
- Verapamil (athletic individual)
- Flecainide (athletic individual)
- ACE inhibitor if there is LV dysfunction
- Catheter ablation (in symptomatic patients who have failed medical therapy ± have evidence of ventricular dysfunction).

RVOT PVCs

Ventricular ectopy associated with a structurally normal heart most commonly occurs from the right ventricular outflow tract. These arrhythmias may occur spontaneously at rest or are induced by exercise at a critical rate, in the recovery phase of exercise, by isoprenaline, or related to the hormonal changes of pregnancy, menses, and menopause. The characteristic ECG pattern for these arrhythmias is a large, tall R wave in the inferior leads (inferior axis) with a left bundle-branch block pattern in V1 (deep pure S wave and no secondary R wave). Beta-blocker therapy is first-line therapy if symptomatic.

Patients with RVOT PVCs and reduced cardiac function, or those with proven RVOT VT may benefit from catheter ablation.

Contributor: Orhan Uzun

6.26 Rheumatic Fever

Clinical Manifestations (Revised Jones Criteria 2015)

The modified Jones criteria (2015)

LR populations

Major criteria	Minor criteria
1. Carditis Clinical and/or subclinical 2. Arthritis Polyarthritis only 3. Chorea 4. Subcutaneous nodules 5. Erythema marginatum	1. Polyarthalgia 2. Fever (≥38.5°C) 3. ESR ≥ 60 mm/h and/or CRP ≥3.0 mg/dL 4. Prolonged PR interval after accounting for age variability

Moderate risk to HR populations

Major criteria	Minor criteria
1. Carditis Clinical and/or subclinical 2. Arthritis Monoarthritis or polyarthritis Polyarthralgia 3. Chorea 4. Subcutaneous nodules 5. Erythema marginatum	1. Monoarthralgia 2. Fever (≥38°C) 3. ESR ≥30 mm/h and/or CRP ≥3.0 mg/dL 4. Prolonged PR interval after accounting for age variability

CRP, C-reactive protein; *ESR*, erythrocyte sedimentation rate.
For all patient populations with evidence of preceding Group A streptococcal infection.
Diagnosis of initial ARF: 2 major manifestations or 1 major plus 2 minor manifestations.

6.26.1 Investigations

- Confirm diagnosis with 2 major **OR** 1 major and 2 minor manifestations **PLUS** evidence of Group A Streptococcal infection
- Detail history including clear documentation of any usage of paracetamol or NSAIDs that could mask the clinical manifestations
- Bloods (WBC, ESR, CRP, consider culture if febrile)
- Baseline ECG and Echo – repeat echo in 1 month if original scan was normal
- Throat swabs, ASOT and anti-DNase titres – repeat test in 10-14 days if first test is not confirmatory

Morphological Findings on Echocardiogram in Rheumatic Valvulitis

Acute mitral valve changes
Annular dilation
Chordal elongation
Chordal rupture resulting in flail leaflet with severe mitral regurgitation
Anterior (or less commonly posterior) leaflet tip prolapse
Beading/nodularity of leaflet tips
Chronic mitral valve changes: not seen in acute carditis
Leaflet thickening
Chordal thickening and fusion
Restricted leaflet motion
Calcification
Aortic valve changes in either acute or chronic carditis
Irregular or focal leaflet thickening
Coaptation defect
Restricted leaflet motion
Leaflet prolapse

Echo findings in Rheumatic Valvulitis

Pathological mitral regurgitation (all 4 criteria met)
Seen in at least 2 views
Jet length ≥2 cm in at least 1 view
Peak velocity >3 m/s
Pansystolic jet in at least 1 envelope
Pathological aortic regurgitation (all 4 criteria met)
Seen in at least 2 views
Jet length ≥1 cm in at least 1 view
Peak velocity >3 m/s
Pan diastolic jet in at least 1 envelope

Reference Range for ASOT and anti-DNase

Age (years)	ASOT (U/ml)	Anti-DNase
1 – 4	170	366
5 – 1	276	499
15 – 24	238	473
25 – 34	177	390
≥ 35	127	265

6.26.2 Management

- Eradicate organism (oral Penicillin V or Erythromycin for 10 days)
- Consider admitting patient to confirm diagnosis, by stopping NSAIDs and observing for clinical symptoms. Start paracetamol or codeine if necessary
- Manage carditis with fluid restriction, diuretics and ACE inhibitors. Consider steroids in severe carditis
- Manage arthritis with Aspirin (anti-inflammatory dose) or NSAIDs

6.26.3 Secondary Prophylaxis of Rheumatic Fever (Recurrent Attacks)

Agent	Dose	Mode
Benzathine Penicillin G	≥ 20kg : 900mg (1,200,000 U) deep IM 4 weekly < 20kg : 450mg (600,000 U) deep IM 4 weekly	IM
Penicillin V	250 mg twice daily	Oral
Erythromycin	250 mg twice daily	Oral

6.26.4 Duration of Secondary Rheumatic Fever Prophylaxis

Manifestation	Treatment
Uncertain ARF	Secondary prophylaxis for 12 months, or until an alternative diagnosis is confirmed
Highly suspicious ARF	10 years or until age 21 (whichever is longer)

Status after initial period elapsed

Manifestation	Treatment
No or Mild RHD	Discontinue treatment
Moderate RHD	Continue until 35 years of age
Severe RHD	Continue until 40 years of age, or longer

References:

1) Revision of the Jones Criteria for the Diagnosis of Acute Rheumatic Fever in the Era of Doppler Echocardiography: A Scientific Statement From the American Heart Association. Circulation. 2015;131:000-000. DOI: 10.1161/CIR.0000000000000205.

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Contributors: Amos Wong and Sandeep Ashtekar (due for review 2026).

6.27 RSV Infection in Cardiac Patients

Respiratory syncytial virus is the most common cause of **bronchiolitis** and pneumonia among infants and children under 1 year of age. The illness generally begins with coryzal symptoms, progressing to cough and sometimes wheezing. During their first RSV infection, between 25% and 40% of infants and young children have signs or symptoms of bronchiolitis or pneumonia, and 0.5% to 3% require hospitalization. Most children recover from illness in 8 to 15 days. The illness can be severe in cardiac babies, particularly those with L→R shunt or with pulmonary hypertension. The mortality in high risk patients is ~3%.

The Joint Committee for Vaccinations and Immunisation recommended that the following should receive passive immunization with palivizumab:
https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/458469/Green_Book_Chapter_27a_v2_0W.PDF

High Risk due to Congenital Heart Disease (CHD)

a) Preterm infants with haemodynamically significant, acyanotic CHD at the chronological ages at the start of the RSV season and gestational ages at birth covered within the **light** green shaded area in Table 1 (below).

b) Cyanotic or acyanotic CHD **plus** significant co-morbidities particularly if multiple organ systems are involved.

Table 1 – Cost effective use of Palivizumab (shaded area)

	Gestational age at birth (weeks+days)						
Chronological age (months)	≤24^{+0}	24^{+1} to 26^{+0}	26^{+1} to 28^{+0}	28^{+1} to 30^{+0}	30^{+1} to 32^{+0}	32^{+1} to 34^{+0}	≥34^{+1}
<1.5							
1.5 to <3							
3 to <6							
6 to <9							
≥9							

The committee does not recommend routine RSV prophylaxis for all cardiac babies, but does state:
"Where clinical judgement of other individual patient circumstances strongly suggests that prophylaxis would prevent serious RSV infection in infants who are at particular risk of complications from RSV, use of Synagis® could be considered during the RSV season."

An RSV information sheet is available on the shared directory "Info Sheet" section). It **should be given to the parents of all infants with haemodynamically significant congenital**

heart disease and the precautions apply during the baby's first RSV season (as a minimum).

In patients where Palivizumab us considered to be beneficial, it should be administered in their local health board. Cardiff patients, and those for whom local administration proves impossible, should be referred to the Nurse-Led Palivizumab Clinic (speak to the CNS team).
Contributor: Dirk Wilson – reviewed 2023

6.27.1 Cardiac surgery following respiratory viral infection

Cardiac surgery may need to be delayed in infants who have suffered a recent viral infection. The following is advised:

Virus	Recovery period pre surgery (non-bypass)	Recovery period pre surgery (bypass pump)	Comment
RSV, Human metapneumovirus and Covid-19	4-6 weeks from first symptoms or positive NPA and clinically well	6 weeks from first symptoms or positive NPA and clinically well	Earlier surgery may be justified for clinical reasons
Other viruses, e.g. Influenza, Parainfluenza, Rhinovirus, seasonal Coronavirus	2 weeks from first symptoms or positive NPA/swab and clinically well	4 weeks from first symptoms or positive NPA/swab and clinically well	

Please note that, because viral RNA is now tested, the result may remain positive for 2-4 weeks after the infection has clinically cleared and therefore we should probably not insist on "virus negativity" prior to surgery if the patient is symptomatically well.
Contributors: Shafi Mussa, Andrew Parry, Dirk Wilson – reviewed 2023

6.28 Screening for Cardiac Disease

(genetic, familial, post-chemotherapy, etc)

Causes of familial cardiac disease include:

- Heart muscle disease
  - Arrhythmogenic right ventricular cardiomyopathy (ARVC) / arrhythmogenic cardiomyopathy (AC)
  - Dilated cardiomyopathy (DCM)
  - Duchenne muscular dystrophy and other dystrophin problems
  - Hypertrophic cardiomyopathy
  - LV non-compaction cardiomyopathy
  - Cardiomyopathy related to mitochondrial disease
  - Post-chemotherapy screening
- Heart rhythm problems
  - Brugada syndrome
  - Long QT syndrome (LQTS)
  - Catecholaminergic polymorphic ventricular tachycardia (CVPT)
- Heart ± aorta structural problems
  - Vascular Ehlers-Danlos Syndrome
  - Marfan syndrome
  - Loeys-Dietz syndrome
  - Famililal aortopathy (non-syndromic)

6.28.1 Heart muscle disease

(See *Eur Heart J* 2010;31:2715-2728)

ARVC / AC screening

In first degree relative (parent or sibling) – assume 50% risk of inheriting the gene:

- Ensure involvement of medical genetics; consider predictive testing if available
- Screening interval: 3 yearly in <10s, 1-2 yearly in patients aged 10-20, 2-5 yearly age 20+, stop screening age 50-60
- Full history and examination, including family history
- ECG (looking for epsilon wave in V1), signal averaged ECG (SAECG)
- Echo (looking for RV dilatation, reduced RV function, RV thinning – LV can rarely be involved)
- Consider need for MRI scanning
- Transfer to adult cardiology age 16-18

If features of ARVC noted, or if presymptomatic genetic diagnosis is made:

- For annual risk factor assessment, including Holter monitoring (looking for ventricular arrhythmia) and exercise test (exercise-induced arrhythmias)
- Consider need for EPS/ICD
- Discuss with / transfer to adult cardiology at age 16-18.

ARVC in more distant relative:

- Ensure involvement of medical genetics
- Full history and examination
- ECG, echo
- If normal, it may be appropriate to discharge the patient
- If abnormal features noted, see above – regular screening needed with transfer to adult cardiology beyond age 16-18

Dilated cardiomyopathy screening

DCM in first degree relative (parent or sibling) – assume 50% risk of inheriting the gene, although not all DCM has a genetic origin:

- Ensure involvement of medical genetics; consider predictive testing if available
- Screening interval: laminopathies – annually from birth; 1-3 yearly in <10s, 1-2 yearly in 10-20 year olds, 2-5 yearly in >20s; stop screening age 50-60 years
- Full history and examination, including family history
- ECG (looking for LVH, ST/T changes)
- Holter if sudden cardiac death in the index case
- Echo (looking for increased LV size, or abnormal function on M-mode and tissue Doppler)
- Annual screening, particularly during adolescence
- Discuss with / transfer to adult cardiology at age 16-18.

If features of DCM noted, or if presymptomatic genetic diagnosis is made:

- Holter and exercise test assessment
- Give advice about athletic participation (see separate section)
- Consider the need for drug or device intervention (CRT/ICD)
- Discuss with / transfer to adult cardiology at age 16-18.

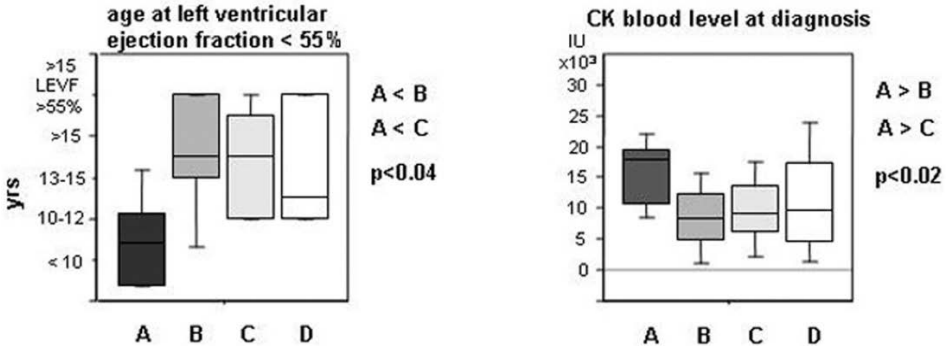
DCM in more distant relative:

- Ensure involvement of medical genetics
- Full history and examination

- ECG, echo
- If normal, it may be appropriate to discharge the patient
- If abnormal features noted, see above – regular screening needed with transfer to adult cardiology beyond age 16-18

Duchenne Muscular Dystrophy Screening

Most boys with DMD will develop cardiac dysfunction as they mature. Use of non-invasive ventilation has improved respiratory outcomes so most deaths are now due to cardiac complications.



"Infantile onset" DMD (group A in figure above) have the following features:

- Early significant motor and intellectual impairment presenting <2 years of age
- High CK at presentation
- Early incidence of cardiac dysfunction

These patients need annual screening from diagnosis.

For other patients with "typical" DMD, he following is a practical approach to screening of myocardial dysfunction in this group of patients:

- Referral for baseline CVS assessment at 4-6 years of age
- Repeat CVS assessment including ECG and echo at loss of ambulation
- 2 yearly assessments if no problem is detected
- After age 10-12, annual assessments depending on the clinical picture; if reduced cardiac function or pulmonary hypertension noted, more frequent follow-up may be needed
- CVS screening should be undertaken before any elective surgery (data <6 months old are acceptable)
- Consider need and feasibility of advanced imaging, e.g. contrast MRI, and testing of pro-NT BNP

Treatment

- The use of an **ACEi or ARB** in the setting of a reduced EF is recommended for all neuromuscular disorders (NMD) *(Class I; Level of Evidence B)*.
- The use of an **ACEi or ARB** before onset of a reduced FS/EF in boys with DMD age ≥10 years may be considered *(Class IIb; Level of Evidence B)*. Discussion with parents about risks/benefits of drug intervention in patients with no evidence of LV dysfunction is needed.
- Given the balance of human data regarding the use of **β-adrenergic blockade** in DMD/BMD and, to a lesser extent, other neuromuscular disorders (NMD), the use of β-adrenergic blockade in the setting of any NMD with a reduced EF is recommended *(Class I; Level of Evidence B)*.

- Without other indication (eg, arrhythmia), the use of β-adrenergic blockade in the absence of reduced EF as therapy to delay or prevent onset of dilated cardiomyopathy is currently not recommended *(Class III; Level of Evidence C)*.
- Given the evidence of benefit in adults with symptomatic LV systolic dysfunction, it is reasonable to consider the use of an **aldosterone antagonist** in DMD/BMD with systolic dysfunction *(Class IIa; Level of Evidence C)*.
- Use of an aldosterone antagonist in DMD/BMD and with preserved LV systolic function, particularly in those who have evidence of myocardial fibrosis (eg, LGE on CMR), may be considered *(Class IIb; Level of Evidence C)*.
- **Corticosteroids** may also offer cardioprotection (these are usually started by the Neurology / Duchenne team)

Discuss with / transfer to adult cardiology at age 16-18.

References: PLOSone 2009;4:e4347 and
http://www.treat-nmd.eu/userfiles/The_diagnosis_and_management_of_DMD_Lancet_complete.pdf
Pediatrics 2005;116 (6):1569-1573
Lancet Neurol. 2014 Dec 29. pii: S1474-4422(14)70318-7
https://www.ahajournals.org/doi/full/10.1161/CIR.0000000000000526

Becker muscular dystrophy

Cardiac dysfunction is rarely encountered <16 years of age.

- Referral for baseline CVS assessment at diagnosis
- Repeat CVS assessment including ECG and echo 3-5 yearly assessments if no problem is detected
- If reduced cardiac function noted, more frequent follow-up may be needed
- Discussion with parents about risks/benefits of drug intervention in patients with no evidence of LV dysfunction; intervention with ACE inhibitors (± β-blocker) if FS <25% or if LVDD > 2 Z-scores from mean;
- Discuss with / transfer to adult cardiology at age 16-18.

References:
Pediatrics 2005;116 (6):1569-1573
Neuromuscular Disorders 2003;13:166-72
https://www.ahajournals.org/doi/full/10.1161/CIR.0000000000000526

Screening of female first-degree relatives of males with Duchenne or Becker muscular dystrophy

- **Known carriers** of the dystrophin gene should be made aware of the risk of developing cardiomyopathy and educated about the signs and symptoms of heart failure (the risk is ~10%). CVS screening with clinical assessment, ECG and echo should take place as follows:
    - At any age if there are symptoms
    - In the absence of symptoms from the age of 16 and 3 to 5 yearly after that depending on the clinical findings.
    - Abnormalities in LV function are more likely to develop after late adolescence and CVS screening needs to continue into adult life.
- Female first-degree relatives of males with Duchenne muscular dystrophy whose **gene status is *not known*** should only undergo CVS screening if there are concerning clinical or laboratory features (e.g. muscle weakness, elevated CK levels). Consideration can be given to screen for CVS abnormalities from late adolescence onwards, although screening is likely to be cost-effective only in known carriers.

Treatment of cardiac disease in carriers is similar to that outlined for boys with DMD or BMD.

References:
Pediatrics 2005;116 (6):1569-1573, Neuromuscular Disorders 2003;13:166-72
https://www.ahajournals.org/doi/full/10.1161/CIR.0000000000000526

Hypertrophic cardiomyopathy screening

HCM in first degree relative (parent or sibling) – assume 50% risk of inheriting the gene:

- Ensure involvement of medical genetics; consider predictive testing if available
- Screening interval: 3-5 yearly before age 10, 1-2 yearly age 10-20, 2-5 yearly after 20; stop screening age 50-60
- Full history and examination, including family history
- ECG (looking for LVH, ST/T changes)
- Echo (looking for septal or apical hypertrophy, function, LVOT velocity)
- Discuss with / transfer to adult cardiology at age 16-18.

If features of HCM noted, or if presymptomatic genetic diagnosis is made:

- Annual risk factor assessment should be undertaken – see https://hcmriskkids.org Parameters assessed are age, gender, weight (kg), LV maximal wall thickness (mm), LA diameter (mm), LVOT gradient (mmHg), NSVT y/n (annual Holter needed), unexplained syncope y/n.
- Consider value of exercise test (looking for abnormal BP response, exercise-induced arrhythmias)
- Give advice about athletic participation (see separate section)
- Consider need for ICD if deemed high risk
- Discuss with / transfer to adult cardiology at age 16-18.

HCM in more distant relative:

- Ensure involvement of medical genetics
- Full history and examination
- ECG, echo
- If normal, it may be appropriate to discharge the patient
- If abnormal features noted, see above – regular screening needed with transfer to adult cardiology beyond age 16-18

LV non-compaction screening

LVNC in first degree relative (parent or sibling) – assume 50% risk of inheriting the gene:

- Ensure involvement of medical genetics; consider predictive testing if available
- Screening interval: start screening from newborn period, screen 1-3 yearly before age 20, then 2-5 yearly age 20+; stop screen age 50-60
- Full history and examination, including family history
- ECG (looking ST/T changes)
- Echo (looking for ventricular function and evidence of spongy/non-compacted area of myocardium)
- Annual screening, particularly during adolescence
- Discuss with / transfer to adult cardiology at age 16-18.

If features of LVNC noted:

- Consider MRI
- Commence standard anti-failure treatment
- Consider need for anti-platelet therapy or formal anticoagulation (potential risk of CVA, but this risk is relatively low if the patient is in sinus rhythm)
- Give advice about athletic participation (see separate section)
- Consider need for biventricular pacing or ICD
- Transfer to adult cardiology at age 16-18.

LVNC in more distant relative:

- Ensure involvement of medical genetics
- Full history and examination
- ECG, echo

- If normal, it may be appropriate to discharge the patient
- If abnormal features noted, see above – regular screening needed with transfer to adult cardiology beyond age 16-18

Screening post chemotherapy exposure
The International Late Effects of Childhood Cancer Guideline Harmonization Group Lancet Oncology 2023 has proposed the following for patients who had the following treatments for childhood cancer:

- Daunorubicin
- Doxorubicin
- Epirubicin
- Mitozantrone
- Idarubacin
- Amsacrine
- ?High dose cyclophosphamide
- Radiotherapy to thorax, thoracic spine, or mediastinum (including left flank and total body irradiation)

Risk stratification:
Anthracycline

Cardiomyopathy surveillance is recommended for survivors of CAYA cancer treated with high doses (≥250 mg/m²) of anthracyclines (high-quality evidence, strong recommendation)
Cardiomyopathy surveillance is reasonable for survivors of CAYA cancer treated with moderate doses (≥100 to <250 mg/m²) of anthracyclines (high-quality evidence, moderate recommendation)
Cardiomyopathy surveillance is not recommended for survivors of CAYA cancer treated with low doses (<100 mg/m²) of anthracyclines (high-quality evidence, strong recommendation)

Chest radiotherapy

Cardiomyopathy surveillance is recommended for survivors of CAYA cancer treated with high doses (≥30 Gy) of chest-directed radiotherapy (high-quality evidence, strong recommendation)
Cardiomyopathy surveillance is reasonable for survivors of CAYA cancer treated with moderate doses (≥15 to <30 Gy) of chest-directed radiotherapy (high-quality evidence, moderate recommendation)
Cardiomyopathy surveillance is not recommended for survivors of CAYA cancer treated with low doses (<15 Gy) of chest-directed radiotherapy with conventional fractionation (high-quality evidence, strong recommendation)

Combination therapy:

Cardiomyopathy surveillance is recommended for survivors of CAYA cancer treated with moderate-to-high doses of anthracyclines (≥100 mg/m²) and moderate-to-high doses of chest-directed radiotherapy (≥15 Gy; high-quality evidence, strong recommendation)

Recommended surveillance:

High risk patients:

Cardiomyopathy surveillance is recommended for survivors of CAYA cancer at high risk to begin no later than 2 years after the completion of cardiotoxic therapy and to continue every 2 years thereafter (moderate-quality evidence, strong recommendation)
Lifelong cardiomyopathy surveillance is reasonable for survivors of CAYA cancer at high risk (expert opinion, moderate recommendation)

Moderate risk patients:

Cardiomyopathy surveillance is reasonable for survivors of CAYA cancer at moderate risk to begin no later than 2 years after the completion of cardiotoxic therapy, to be repeated at 5 years after diagnosis, and to continue every 5 years thereafter (low-quality evidence, moderate recommendation)
Lifelong cardiomyopathy surveillance is reasonable for survivors of CAYA cancer at moderate risk (expert opinion, moderate recommendation)

Low risk patients:

Cardiomyopathy surveillance is not recommended for survivors of CAYA cancer at low risk (moderate-quality evidence)

When abnormalities are found:

Cardiology consultation is recommended for survivors of CAYA cancer with asymptomatic left-ventricular systolic or diastolic dysfunction* following treatment with anthracyclines or chest-directed radiotherapy (expert opinion, strong recommendations)
Treatment with heart failure medications (eg, ACE inhibitors, ARBs, or β blockers) is recommended in survivors of CAYA cancer with asymptomatic left-ventricular ejection fraction <40%, according to guidelines from the general population (low-quality to high-quality evidence in the general population, strong recommendation)
No recommendations can be formulated about treatment with heart failure medications in survivors of CAYA cancer with asymptomatic borderline cardiac function (left-ventricular ejection fraction between 40% and the lower limit of typical function; no studies in survivors of CAYA cancer and no evidence in the general population)

Transition to adult services:

- **Low risk patients**
  Will not be having regular surveillance – no need to transfer care.
- **Moderate risk patients**

Discharge to GP/late effects clinic age 16-18 years– advise 5 yearly and pre-pregnancy CVS assessment.

- **High risk patients**
  Transfer to local adult cardiac services age 16-18 years

Reference: Systematic review and updated recommendations for cardiomyopathy surveillance for survivors of childhood, adolescent, and young adult cancer from the International Late Effects of Childhood Cancer Guideline Harmonization Group
Lancet Oncology 2023

6.28.2 Heart Rhythm

See https://academic.oup.com/europace/article/24/8/1307/6562982

Brugada Syndrome Screening
In first degree relative (assume 50% risk):

- Ensure involvement of medical genetics; consider predictive testing if available
- Full history and examination, including family history
- ECG (looking for characteristic changes)
- Echo (to rule out features of ARVC – the ECG changes can overlap and there may be some diagnostic confusion)
- Annual clinical and ECG surveillance
- Give generic "Brugada advice"
  - Drug avoidance (Brugadadrugs.org)
  - Fever management
  - Avoid carbohydrate loading before sleep
  - Electrolyte replacement with athletic participation
- Discuss with / transfer to adult cardiology at age 16-18

Clinical suspicion or if presymptomatic genetic diagnosis made:

- Consider need for signal averaged ECG (may be helpful in diagnosis and risk stratification)
- Consider need for ajmaline/flecainide challenge in children, usually >16 years of age (mandatory at any age if symptomatic)
- Consider need for VStim study (if +ve ECG or drug challenge, or if symptomatic)
- Consider need for ICD
- 6 monthly review
- Give specific "Brugada advice"
  - Drug avoidance (Brugadadrugs.org)
  - Fever management
  - Avoid carbohydrate loading before sleep
  - Electrolyte replacement with athletic participation, which is permitted (see https://www.escardio.org/Journals/E-Journal-of-Cardiology-Practice/Volume-19/brugada-syndrome-and-sports-activity-from-history-to-risk-stratification#:~:text=Malignant%20arrhythmias%20in%20Brugada%20syndrome,tone%20may%20promote%20nocturnal%20arrhythmias.)
- Discuss with / transfer to adult cardiology at age 16-18.

Brugada syndrome in more distant relative:

- Ensure involvement of medical genetics
- Full history and examination
- ECG, consider need for echo
- If assessment is normal, it may be appropriate to discharge the patient

- If abnormal features noted, see above – regular screening needed with transfer to adult cardiology beyond age 16-18

CPVT Screening

CPVT is characterized by episodic syncope occurring during exercise or acute emotion in individuals without structural cardiac abnormalities. The underlying cause of these episodes is the onset of fast ventricular tachycardia (typically bidirectional or polymorphic). Spontaneous recovery occurs when these arrhythmias self-terminate. In other instances, ventricular tachycardia may degenerate into ventricular fibrillation and cause sudden death if cardiopulmonary resuscitation is not readily available. The mean age of onset of CPVT is between seven and nine years; onset as late as the fourth decade of life has been reported. It has been associated with two genes that make proteins found inside the cell – the human ryanodine receptor (a calcium ion channel) and calsequestrin (a protein that interacts with the channel). These regulate the release of calcium ions into the rest of the cell. If these do not function normally, the level of calcium inside the cell becomes too high, resulting in the arrhythmias characteristic of CPVT.

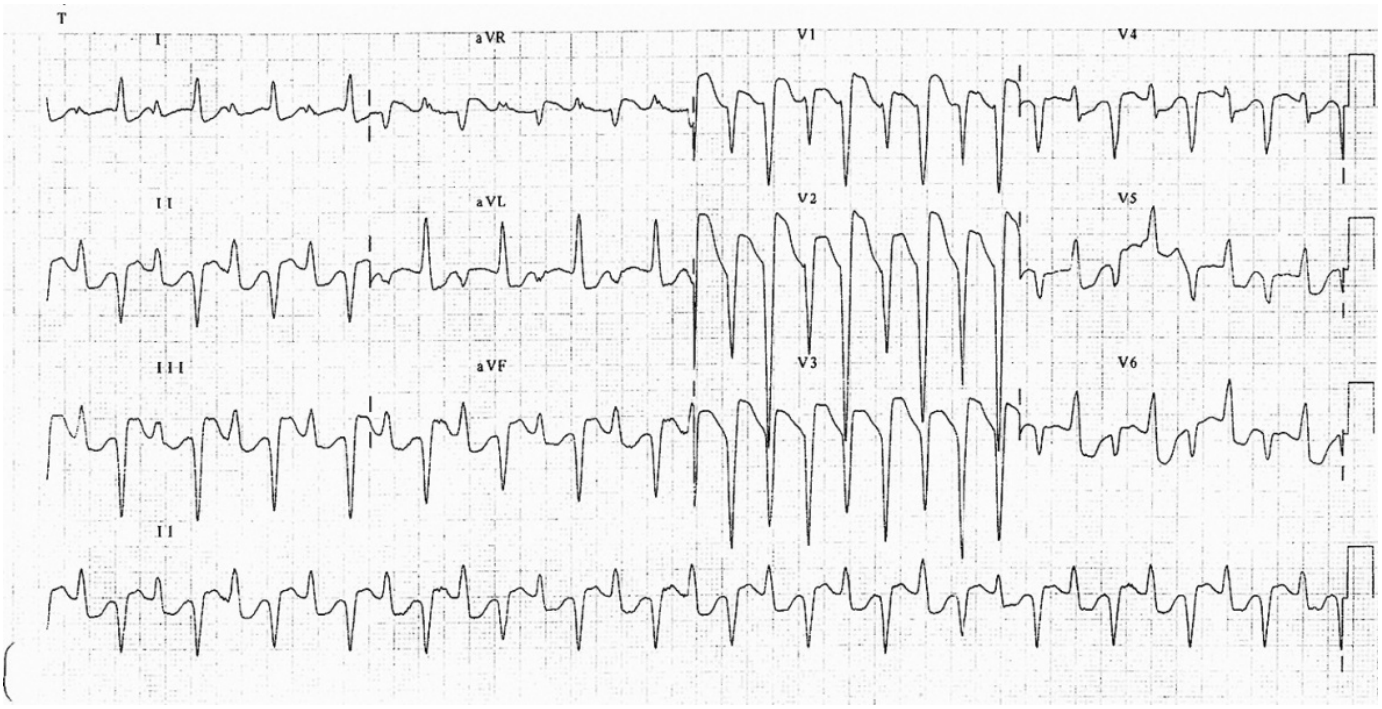


Image: http://lifeinthefastlane.com/ecg-library/basics/bvt/ Typical bidirectional VT seen in CPVT

CPVT in first degree relative (assume 50% risk)

- Ensure involvement of medical genetics; consider predictive testing if available
- ECG
- Holter
- Exercise test
- If abnormal features noted, give advice about exercise restriction (see separate section), prescribe β-blockers, consider need for ICD device
- Discuss with / transfer to adult cardiology at age 16-18.

CPVT in more distant relative

- Ensure involvement of medical genetics
- Full history and examination

- ECG
- Consider need for Holter and exercise test
- Discuss with genetics / adult cardiology about value of screening beyond 16-18 years

Long QT Syndrome Screening
LQTS in first degree relative (assume 50% risk):

- Ensure involvement of medical genetics
- Full history and examination, including family history
- ECG (looking for characteristic changes), consider echo
- Consider need for epinephrine challenge in children > age 8-10 (mandatory if symptomatic)
- Consider need for VStim study (if +ve ECG or drug challenge, or if symptomatic)
- Consider need for intervention (β-blockers, ICD ± pacing)
- Annual screening, but if features of LQTS syndrome noted, or if presymptomatic genetic diagnosis is made, for 6 monthly review
- Give advice regarding athletic participation
- Discuss with / transfer to adult cardiology at age 16-18.

Long QT syndrome in more distant relative:

- Ensure involvement of medical genetics
- Full history and examination
- ECG
- If normal, it may be appropriate to discharge the patient
- If abnormal features noted, see above – regular screening needed with transfer to adult cardiology beyond age 16-18

See https://academic.oup.com/eurheartj/article/43/40/3997/6675633

6.28.3 Heart Structure

Ehlers Danlos Syndrome Screening
EDS is a condition characterized by the following features:

- Joint hypermobility
- Skin hyperextensibility
- Tissue fragility

13 subtypes of EDS are described and a genetic basis is the cause in most of these sub-types (see https://www.ehlers-danlos.com/eds-types/)

The majority of patients with a label of “EDS” have benign hypermobility, which is often familial, but which has no genetic basis. These patients almost never have concerning cardiac pathology. A small subset of patients with hypermobility suffer from joint pain and fatigue; where the Beighton score is 6 or above a label of hypermobility-type EDS (hEDS) may be made – see the RCPCH statement from 2019:
https://www.rcpch.ac.uk/resources/establishing-correct-diagnosis-ehlers-danlos-syndrome-hypermobility-type-heds-children

There is no indication for an individual with benign hypermobility and a normal CVS exam to undergo cardiac surveillance.

If an individual meets the criteria described above for hEDS it is reasonable to undertake a one-off echocardiogram to assess the mitral valve and aortic root. In the absence of a family

history of aortopathy, ongoing surveillance is unlikely to be clinically valuable, i.e. if the FHx and echo are normal the patient can be discharged.

In patients with other forms of genetically proven, but benign forms of EDS may rarely have mitral valve prolapse and regurgitation, or mild aortic enlargement, but intervention is seldom needed. Referral for echo assessment is reasonable, but long-term CVS surveillance is rarely indicated unless these findings are present.

Vascular EDS (COL3A1 mutation) is a rare autosomal dominant condition with a high cardiovascular mortality due to vessel rupture. Children of adults with vascular EDS need to be assessed by medical genetics and if the condition is confirmed in the child, they should undergo repeated cardiovascular assessment through to adult life. Surgical and vascular interventions and pregnancy carry a high morbidity/mortality due to bleeding, vessel rupture and poor wound healing. Attention should be paid to ensuring blood pressure is normal, but there is no specific therapy that is known to reduce the complication rate. The use of the beta-blocker Celiprolol is advocated by some (Lancet 2010;376(9751):1476-84).

Marfan Syndrome Screening
If MFS in first degree relative (assume 50% risk):

- Refer to paediatric Marfan syndrome clinic
- Full history and clinical examination
- ECG, echo (aortic enlargement, mitral or tricuspid valve prolapse, MR)
- Consider need for fibrillin mutation analysis
- 6-12 monthly review
- Consider need for β-blockers
- Give advice about athletic participation, contraception, pregnancy (where relevant)
- Transfer to GUCH service age 16-18

Marfan clinic referral criteria
The purpose of the joint Paediatric Marfan/Aortopathy Clinic is for the assessment or management of children (<18 years old) with:

1. Confirmed Marfan / other aortopathy condition (e.g. Loeys Dietz and vascular EDS cases (by clinical or genetic diagnosis)
2. Individuals at 1 in 2 risk of Marfan / other aortopathy in known families where genetic testing has not been informative or the patient has declined testing
3. Dilated aorta (Z score ≥2) with an as yet unknown genetic cause
4. Patients with dislocated or subluxed lenses with Marfan as a possible diagnosis

Outside the situations above, the clinic is not for general referrals on children with a queried diagnosis of Marfan syndrome. These should go to the locality genetic counsellor / consultant team, and will require a full genetic counsellor work up and consultant appointment. Echo assessment may form part of the assessment – this can be performed by the local paediatrician with expertise in cardiology.

Turner Syndrome Screening
50% of Turner patients have aortic enlargement, irrespective of presence or absence of a bicuspid aortic valve (Cardiol Young 2009;19:568-72). The risk of aortic dissection is 6x the general population (but is lower than Marfan patients). Best practise:

- Full history and clinical examination
- Detailed echo at diagnosis – look for bicuspid aortic valve and coarctation; assess and document aortic z-scores (A1-4, as with Marfan and other aortopathies)

- Manage BAV/CoA as clinically appropriate (if present) – continue to include A1-A4 assessment as part of long-term surveillance – NB use the Turner specific z-score calculator – see http://www.parameterz.com/refs/quezada-ajmg-2015
- If there is no BAV/CoA, continue to see the patient (e.g. age 5, age 10, then 1-3 yearly depending on findings with A1-4 assessment
- Consider need for MRI scan age 10-11 and age 16
- Consider β-blockage (or angiotensin receptor blocker) if aorta >97th percentile for size (z-score >2)
- Refer for surgical aortic root replacement if aorta >55 mm in an adult, or >25 mm/m^2 (or aortic z-score >5-6 in a child), or if the aorta is rapidly increasing in size

See also http://adc.bmj.com/content/early/2015/01/08/archdischild-2014-307080.full.pdf+html?sid=aacaa483-5069-4aca-98eb-42e1c9189936

https://www.ahajournals.org/doi/full/10.1161/HCG.0000000000000048

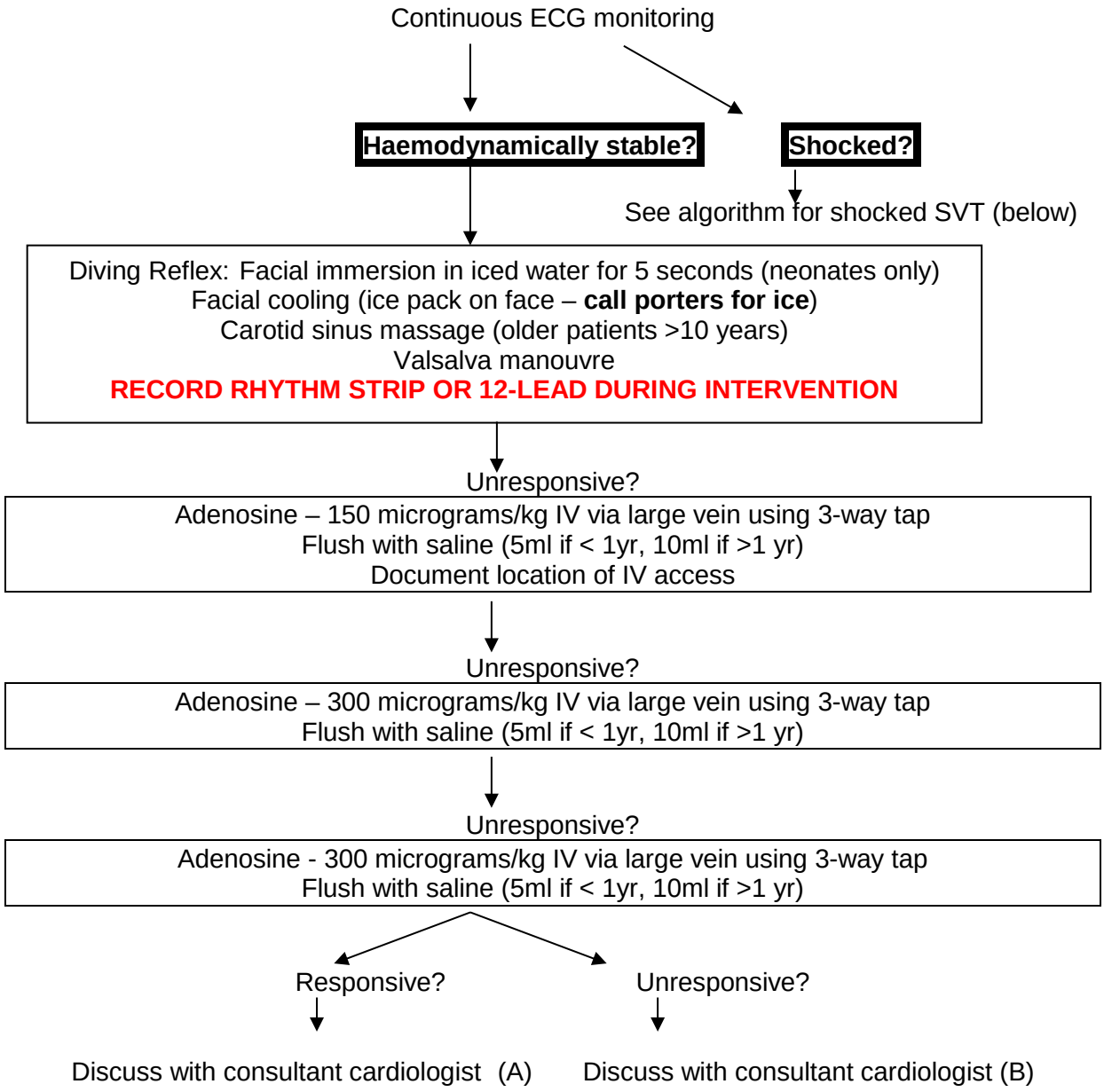
Contributors: Sandeep Ashtekar, Orhan Uzun, Dirk Wilson – guideline reviewed 2023

6.29 Supraventricular Tachycardia

Diagnosis

- Inappropriate tachycardia (usually > 180-220/min)
- Narrow QRS complex
- No beat-to-beat variation
- Absent/abnormal P-waves

Treatment algorithm - Inform relevant consultant



A – Infants <12 months – check capillary blood gas. If blood gas and ventricular function normal, see "SVT Prophylaxis" section below. If ventricular function is not known, digoxin may be a safer acute option.

B – Infants <12 months – check capillary blood gas. Check ventricular function with echo. If blood gas and/or function are abnormal, transfer to HDU/PICU environment for continuous monitoring. Consider DC cardioversion or digoxin loading dose or careful amiodarone infusion with inotropic support (see Amiodarone in Therapeutics section). If blood gas/ventricular function normal, consider oral flecainide 1-3 mg/kg stat or IV digoxin or cautious IV amiodarone (see Therepeutics section). If still in SVT 30 mins after giving antiarrhythmic, give further dose of adenosine, 300 microgram/kg.

SVT – haemodynamically unstable (Shocked SVT)

↓

Oxygen, Airway, Breathing

CONSIDER NEED FOR SEDATION OR GENERAL ANAESTHESIA

Synchronous DC Cardioversion, 1 J/kg

↓ Unresponsive?

Synchronous DC Cardioversion, 2 J/kg

↓ Unresponsive?

Consider IV Sodium Bicarbonate between shocks

Responsive? ↓ Start prophylaxis

↓ Unresponsive? Discuss with cardiologist, consider IV amiodarone (5 mg/kg over 60-120 min) see Therapeutics section for advice on dosage

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SVT Prophylaxis

ECG

WPW

Check no obstructive heart lesion, ensure good cardiac function

Flecainide 1 to 3 mg/kg/dose twice daily (safest to start as an inpatient and check ECG the next day – rule out Brugada pattern)

No WPW

<1 yr: Flecainide 2 to 3 mg/kg/dose twice daily or Propranolol 0.25 to 1 mg/kg/dose tds

>1 yr: Atenolol 1 mg/kg/dose od, or 0.5 to 1 mg/kg/dose bd, or Consider flecainide (see below)

Trough flecainide levels (at least 6 hours post-dose, e.g. dose given 8 AM, check level 2 pm). Consider toxicity if level is elevated or if QRS duration increases >25% from baseline. **See Section 7.8 – Flecainide for SVT.**

Some individuals may require combination therapies. Effective combinations are:

Flecainide – Atenolol (or propranolol)
Flecainide – Digoxin (may need to reduce digoxin dose)
Sotalol – Digoxin
Atenolol (or propranolol) – Digoxin
Atenolol – Propafenone

Where combination treatment is used, watch for ECG changes and monitor with Holter.

6.29.1 WPW Management

A subset of children with WPW are at risk of life-threatening events (LTE). In some individuals, an LTE may be the first presentation of WPW. The risk factors for LTE in children are:

- Pathway capable of rapid conduction (antegrade conduction RR interval cycle length <250 msec) as assessed in EP study
- Atrial fibrillilation inducible at EP study
- Male sex
- Presence of congenital heart disease, especially Ebstein
- Presence of multiple pathways (also associated with Ebstein)

Patients with symptoms of SVT ar less likely to have LTE. Intermittent pre-excitation and disappearance of the pathway on exercise testing might not offer reassurance that the patient is protected from LTE.
See: http://electrophysiology.onlinejacc.org/content/4/4/433

Current unit policy is**:**

- Counsel parents that a small proportion of patients with WPW can have LTE, and that the highest risk period is age 15-30 years and that LTEs are unlikely to (but can) occur below age 8 years
- At present there is no evidence that giving regular medication reduces the risk of LTE
- Manage WPW patients with intermittent palpitations with anti-arrhythmic drugs, usually flecainide as a single agent or combined with another anti-arrhythmic
- Refer any patient with WPW and an LTE or syncope for urgent EP study
- Refer all patients 8+ for EP study

Contributors: Orhan Uzun, Dirk Wilson – guideline reviewed 2023

6.30 Transplantation

Prior to transplantation, patients invariably require a formal transplant assessment, usually in the transplant centre over a period of several days, either in Great Ormond Street Hospital or Freeman Hospital in Newcastle. This usually answers the following questions:

- Is heart transplant possible
- Is it the best treatment available
- Is it the right option for the child at this time

See https://www.gosh.nhs.uk/conditions-and-treatments/conditions-we-treat/heart-transplant/

The main medical considerations after transplantation are immunosupression (maintenance therapy and monitoring of drug levels), prevention and treatment of opportunistic infections and detection of rejection. Close liaison with the transplant centre is essential.

Routine Investigations

When reviewing a patient check the following:

- BP
- Urinalysis
- Weight
- FBC
- U&E, creatinine, LFT
- Trough ciclosporin level
- ECG summated voltages (add peak to peak (R-S or Q-R) voltages in leads I, II, III, V1 & V6).
- Echocardiogram (the need for this reduces with time - liaise with the consultant). Record measurements on the flow sheet in the case record notes and communicate significant changes to the transplant centre.

Immunosupression
Tacrolimus or Neoral (ciclosporin A) and mycophenolate mofetil or azathioprine are the most commonly prescribed drugs.

Ciclosporin (cyclosporin), a calcineurin inhibitor, is a potent immunosuppressant which is virtually non-myelotoxic but markedly nephrotoxic. NB It has multiple drug interactions.

Tacrolimus is a calcineurin inhibitor. Although not chemically related to ciclosporin it has a similar mode of action and side-effects, but the incidence of neurotoxicity and nephrotoxicity appears to be greater; cardiomyopathy has also been reported. Disturbance of glucose metabolism also appears to be significant; hypertrichosis appears to be less of a problem than with ciclosporin.

Azathioprine is widely used for transplant recipients and it is also used to treat a number of auto-immune conditions, usually when corticosteroid therapy alone provides inadequate control. It is metabolised to mercaptopurine, and doses should be reduced when allopurinol is given concurrently. Blood tests and monitoring for signs of myelosuppression are essential in long-term treatment with azathioprine.

Mycophenolate mofetil (MMF) is metabolised to mycophenolic acid which has a more selective mode of action than azathioprine. It is licensed for the prophylaxis of acute rejection in renal or cardiac transplantation when used in combination with ciclosporin and corticosteroids. There is evidence that compared with similar regimens incorporating azathioprine, mycophenolate mofetil reduces the risk of acute rejection episodes; the risk of opportunistic infections (particularly due to tissue-invasive cytomegalovirus) and the occurrence of blood disorders such as leucopenia may be higher.

mTOR inhibitors (sirolimus and everolimus) – Not used routinely. May be used in situations such as cardiac allograft vasculopathy or when there is need to reduce levels of calcineurin inhibitors due to renal dysfunction.

Drug levels and FBC are used to monitor efficacy of immunosupression. Be guided by the transplant centre.

General advice
Immunisations **-** Live vaccines should not be used. Inactive vaccines are suitable (e.g. Salk polio vaccine). Siblings should receive all immunisations including MMR.
Chicken pox - If a patient has significant *exposure* to chicken pox, Zoster immunoglobulin (HZIG) should be given (ideally within 72 hours of exposure). This is reserved for patients on steroids or within a year of transplant. All others receive prophylactic oral acyclovir. Liaise with the PHLS Virologists and the transplant centre.

For *active infection*, give intravenous aciclovir, 10 mg/kg dose three times a day or high dose oral acyclovir, 800 mg five times a day for a total of 10 days.
Endocarditis prophylaxis **-** NICE guidance should be followed (not given routinely).
Diet should be as normal as possible but the following should be avoided due to the risk of bacterial colonisation: undercooked chicken or turkey, shellfish, raw eggs, live yoghurt, blue or soft cheeses, sheep or goats' milk, unpasteurised milk or cream, and possibly pre-prepared salads. Avoid grapefruit juice as mentioned above.

Transplant Rejection
Symptoms/Signs:

- Fever, malaise, anorexia, vomiting
- Breathlessness
- Hepatosplenomegaly
- Gallop rhythm

Investigations:

- Raised WBC, CRP
- ECG changes such as 25% or more reduction in ECG summated voltages, as well as alterations in baseline cardiac rhythm – increased resting heart rate, bradycardia and arrhythmias
- CXR may show cardiomegaly, pulmonary edema or pleural effusion
- Echocardiogram changes include abnormal LV diastolic function (early sign), decreased LV systolic function i.e ↓fractional shortening, ejection fraction (late sign). ↑LVDD and dimensions of LVPW and IVS, new pericardial effusion and new onset AV valve regurgitation
- Endomyocardial biopsy may show histological changes and can be graded according to severity

Treatment: Liaise with transplant team

- Mild rejection - prednisolone 1 mg/kg/day
- Acute severe rejection - intravenous Methylprednisolone. Monitor BP every 15 minutes for first hour. Give Lansoprazole to prevent gastric ulceration. High dose steroids increase Ciclosporin levels

Opportunistic infections

Organism	Prevention	Treatment
Candida or aspergillus	nystatin	amphotericin (caution – renal impairment), ambisome fluconazole
Pneumocystis	co-trimoxazole	high dose intravenous co-trimoxazole
Herpes simplex	oral aciclovir	intravenous aciclovir
CMV	none available	intravenous ganciclovir

Transplant Vasculopathy
This is the leading cause of late death. Risk factors are:

- Older age at transplantation
- Increased number of rejection episodes
- CMV infection
- Hyperlipidaemia.

This may present with graft dysfunction, syncope or it may be asymptomatic until it presents with sudden death.
Coronary angiography is gold standard for diagnosis, and most centres now offer this annually.

Medical treatment involves use of sirolimus and statins, but in most cases re-transplantation is required.

Lymphoproliferative Disease
EBV-related lymphoproliferative disease is a serious and not uncommon (20%) complication of paediatric cardiac transplantation. It presents with non-specific lymphadenopathy. Tonsillar enlargement is not uncommon. There is a UKCCSG protocol for the investigation and management of this disorder – liaise with the local oncology team and the transplant centre.
Contributors: Victor Ofoe, Dirk Wilson – guideline reviewed 2023

6.31 Vaccination / Immunisation in Paediatric Cardiac Patients

Most patients should receive the standard UK immunisation schedule. Special advice is given for asplenia and DiGeorge patients (see section 6.2 and the Green Book https://www.gov.uk/government/collections/immunisation-against-infectious-disease-the-green-book).

6.31.1 Pneumococcal vaccination in paediatric cardiac patients

Any patient with heart disease who would be eligible for the flu vaccine is considered to be at increased risk of pneumoccal infection and they qualify for additional pneumococcal protection. Unless there is a contraindication the standard immunisation schedule should be followed PLUS (see table below):

Age	Vaccine
2 years	*PPV23* – single dose (At least 8 weeks after PCV13 booster)
10 years	*PPV23* – single dose
65 years	*PPV23* – single dose

6.31.2 Cardiac surgery and vaccination

For most paitents All other paediatric cardiac patients should undergo the standard childhood vaccination schedule, however the timing of surgery may lead to delays in the infant vaccination schedule. It is recommended that routine vaccination should be avoided within 2 weeks (and if possible 4 weeks) before cardiac surgery. This will help to prevent added stress during this time period, and will avoid the potential for confusion if a fever develops.

Immunisations should be withheld 2-4 weeks following cardiac surgery. Live vaccines, e.g. MMR, should be withheld for 6 months following bypass surgery or any other blood transfusions. This does not apply to Rotavirus which can be given using the routine schedule.

References:
American Academy of Pediatrics. Kimberlin DW, Brady MT, Jackson MA, Long SS, eds. Red Book: 2015 report of the Committee on Infectious Diseases. 30th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2015
https://www.childrens.health.qld.gov.au/fact-sheet-going-home-after-surgery/
https://starship.org.nz/guidelines/immunisations-and-cardiac-infants/
Guideline reviewed 2023

6.31.3 Flu vaccination and pneumococcal in paediatric cardiac patients

From 2013 Joint Committee on Vaccination and Immunisation (JCVI) summary provides advice on the number of doses of Flu vaccine that children should be offered depending on:

- their age
- whether they have received influenza vaccine before

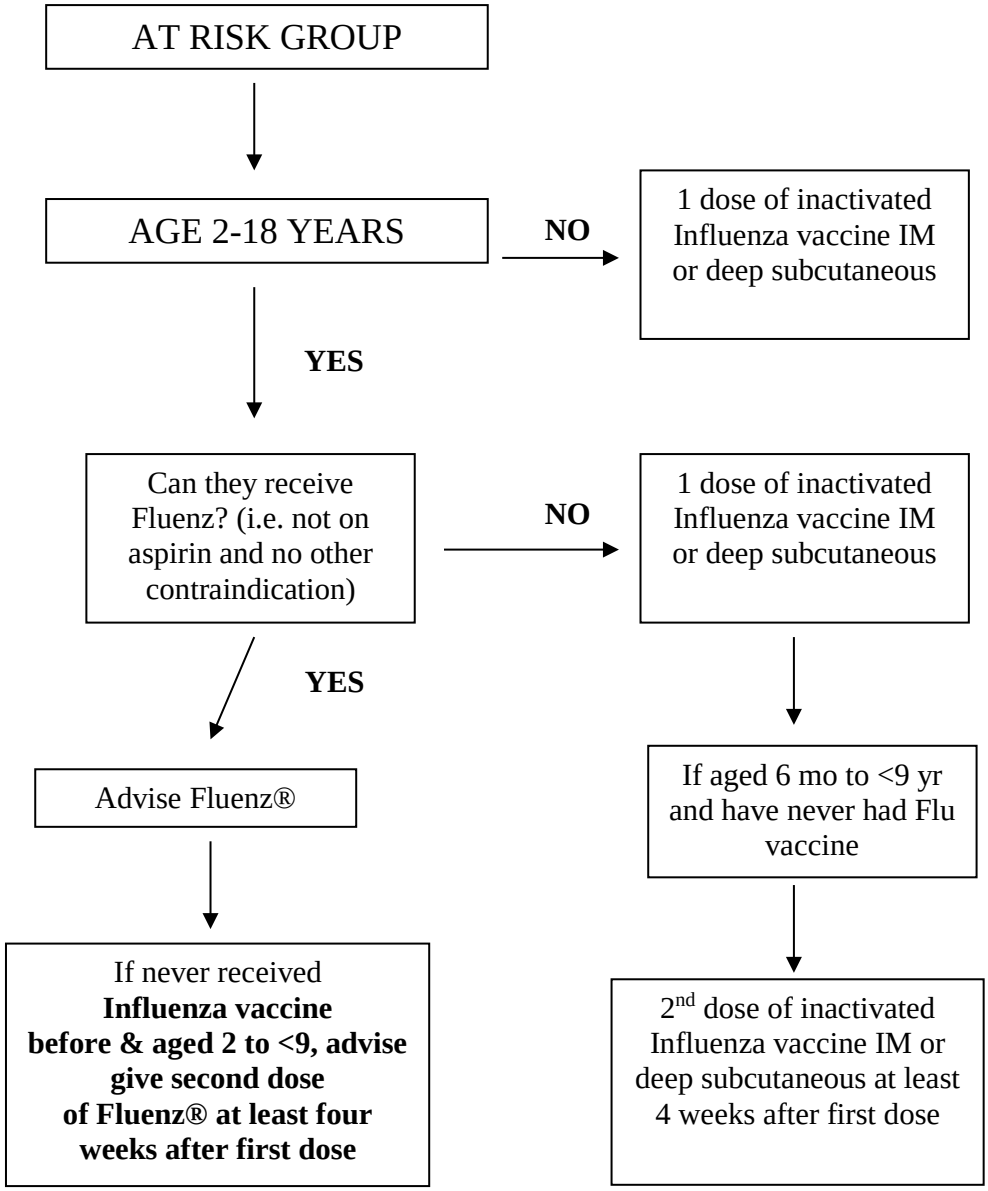
- the type of Flu vaccine

Flu vaccine is recommended for patients over 6 months with:

- Congenital heart disease that is considered haemodynamically significant (severe stenosis or regurgitation of a valve, chamber enlargement, significant ventricular dysfunction, single ventricle anatomy or physiology)
- Hypertension with cardiac complications (e.g. needing medication, evidence of LVH)
- Chronic heart failure
- Those on long-term heart medications
- Ischaemic heart damage

6 months – 2 years in a clinical risk group should have the injected inactivated QIVe. For egg allergic children offer QIVc 'off-label'. Children in this age group who have never had a flu vaccination will need 2 doses 4 weeks apart.

Children ≥ 2 years and not on aspirin can have Fluenz (nasal vaccine).
Flu vaccine and aspirin:
Patients on aspirin should have the inactivated Influenza vaccine intramuscularly (or deep subcutaneous), not live intranasal vaccine (Fluenz). The Influenza vaccine algorithm is shown below:



Source: https://www.gov.uk/government/publications/which-flu-vaccine-should-children-have/flu-vaccines-for-children-and-young-people

Contributors: Claire Logan, Wendy Williams, Dirk Wilson – guideline reviewed 2023
See up-to-date network guidance at https://www.swswchd.co.uk/en/page/clinical-information-children

6.31.4 Prolonged QT syndrome and anaphylaxis from vaccination or other trigger

In cases of prolonged QT syndrome and anaphylaxis the current expert opinion is that it is best to give IM adrenaline as normal (e.g. Epipen) as the risk of death from anaphylaxis outweighs any potential harm. Ref: Correspondence with Dr A Fox and Dr G de Toit Nov 2014.

Contributors: Dr D Tuthill and O Uzun. Guideline reviewed 2019

6.31.5 Varicella zoster virus vaccination

This should be considered in patients who will be on long-term aspirin (to reduce the risk of Reye's syndrome).

SECTION 7 – THERAPEUTICS

7.1 Prescribing Drugs Safely

7.1.1 Medication Errors

Medication errors or serious drug reactions that result in a change in the clinical status of the patient should be dealt with in the following manner:

1. Record the event in the case record notes
2. Discuss the event with the appropriate consultant
3. Discuss the event with the relatives (the consultant may choose to do this)

7.1.2 Preventing Medication Errors

Medication errors are common but fortunately adverse clinical consequences are rare. Prescription errors by doctors account for the majority of errors. Therefore it is essential to ensure the following:

1. An accurate drug history is taken. Bear in mind that parents often know doses in mL not mg and they may use different strengths of liquids at home to those kept in hospital. Always remember to ask about medicines bought over the counter and any alternative medicines (e.g. herbal, homeopathic, Chinese medicines)
2. The prescription chart includes **all** relevant information, including the weight of the patient, date of birth, allergies and all required medications.
3. All handwriting must be neat and clearly legible. Use CAPITAL LETTERS and write in black indelible ink (blue does photocopy, but black is preferred).
4. Avoid abbreviations for drug names and amounts (e.g. write "microgram" instead of using the symbol, µ or "mcg"). Approved names should be used where possible. Take great care in prescribing compound preparations (it is sometimes safer to use brand names in this situation).
5. All prescriptions must be signed and dated at the time of writing.
6. Regular prescriptions can be changed once (dose, frequency or route) – enter the date of change and initial the change.
7. Changes to the chart are discussed with the nursing staff and discontinued medications are crossed out.
8. When drugs are prescribed "as required/prn" the prescription must include the indication for drug administration (e.g. "pain"), the interval between doses and the maximum dose in 24 hours.
9. Use ONE chart per patient, but if the number of medications exceeds one chart, label each chart clearly "chart 1 of 2", chart 2 of 2" etc. Charts must be regularly reviewed to condense the number of charts to a minimum. Continuation sheets are not allowed.
10. The re-writing of charts is a common source of errors – ensure ALL relevant information is transcribed, including weight and allergies. The start date of a drug is

the ORIGINAL start date, not the date the chart was rewritten. Have a colleague or nurse double-check any rewritten chart.

11. Prescription charts should be regularly reviewed for tidiness and legibility. Charts must be rewritten whenever the legibility of a drug is compromised, e.g. something spilled on the chart,
12. When a supplementary chart is used (e.g. steroid, anticoagulant, insulin, etc.) the drug must be identified on the main prescription chart, with time of administration documented, and annotated "see accompanying chart".
13. Numbers for dosages must be clear and unambiguous – take particular care over 4s and 9s, for example.
14. Decimal points must not be employed unless unavoidable. If the use is necessary, the decimal point must be precisely marked and, if appropriate, preceded by a **zero (0), e.g. 0.5 mL, not .5 mL.**
15. In cases where the dose is prescribed in units, e.g. heparin and insulin, the dose must be prescribed as **UNITS, not u**, as there is a risk that a "u" may be mistaken for a "0" and leading to a 10X dose error.
16. When prescribing a liquid preparation (injection, oral mixtures etc.), the dose must be in **milligrams / micrograms / nanograms, not mls**, unless it is avoidable to use a volume.
17. On discontinuation of a prescribed drug, the "crossing off" should occur through the prescribing section of the chart **and** through the section of the chart used to record the drug's administration.
18. The weights for *all* children must be recorded on the drug chart in Kg (the policy only asks for children<12yrs).
19. Ensure that changes to the drugs are communicated to the nursing staff (and patient/parents/carers if appropriate.
20. If in any doubt, speak to the Pharmacy Department.

Reference: **Reviewed 2023**

Pharmacy & Medicines Management Intranet - Good Prescribing Principles BPS.pdf - All Documents (sharepoint.com)

See also http://www.gmc-uk.org/guidance/ethical_guidance/14316.asp and https://www.gmc-uk.org/-/media/documents/Prescribing_guidance.pdf_59055247.pdf

7.1.3 Drug Interactions

Drug interactions are not uncommon in paediatric cardiology. Common potential interactions are:

Drug	Interacts with	Comment
Carvedilol	Flecainide	Reduce flecainide doses by at least 30%, check levels
	Digoxin	Reduce digoxin dose by at least 30%, check levels
Ciclosporin	Macrolide antibiotics	Avoid use, or check ciclosporin levels
Clopidogrel	Proton pump inhibitors	These cause reduced anti-platelet effect
Digoxin	Amiodarone	Reduce digoxin dose by at least 30%, check levels
	Macrolides	Avoid use, or reduce digoxin dose by at least 30%, check levels if to be used long-term
	Carvedilol	Reduce digoxin dose by at least 30%, check levels
	Bosentan	Causes lowering of digoxin levels, but not enough to be clinically significant (http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2014257/)
Warfarin	Amiodarone	INR usually increases – check INR level 1-2 days after starting amiodarone
	Antibiotics	Usually increases INR – check INR level 1-2 days after starting antibiotics
	Bosentan	May reduce the INR – check INR level 1-2 days after starting bosentan

Reviewed 2023 – with thanks to Clara Danielsen

7.2 Quick Calculations of Drug Concentrations for Infusions

DESIRED CONCENTRATION	CALCULATION	DRUGS	DOSE RANGE
Mcg/kg/min	**DOSE in MG in a 50mL Syringe**	**Mcg/kg/min**	
0.01mcg/kg/min = 1mL/hr	0.03 X Weight (Kg)	Adrenaline Noradrenaline PGE1 Isoprenaline	0.01 - 1.0mcg/kg/min 0.01 - 0.5mcg/kg/min 0.005 - 0.05mcg/kg/min 0.025 - 1.0mcg/kg/min
0.02 mcg/kg/min =1mL/hr	0.06 X Weight (Kg)	Adrenaline Noradrenaline	0.01 - 1.0mcg/kg/min 0.01 - 0.5mcgkg/min
0.05 mcg/kg/min = 1mL/hr	0.15 X Weight (Kg)	Adrenaline Noradrenaline	0.01 - 1.0mcg/kg/min 0.01 - 0.5mcg/kg/min
0.1mcg/kg/min=1mL/hr (concentrated)	0.3 X Weight (Kg)	Adrenaline Noradrenaline Isoprenaline	0.01 - 1.0mcg/kg/min 0.01 - 0.5mcg/kg/min 0.025 - 1.0mcg/kg/min
0.33mcg/kg/min = 1mL/hr	1.0 X Weight (Kg)	Milrinone	0.33 - 0.99mcg/kg/min
1 mcg/kg/min = 1mL/hr	3 X Weight (Kg)	Nitroprusside Nitroglycerin Midazolam	0.5 - 10mcg/kg/min 0.5 - 10mcg/kg/min 1 - 6mcg/kg/min
2 mcg/kg/min = 1mL/hr	6 X Weight (Kg)	Midazolam	1 - 6mcg/kg/min
5 mcg/kg/min = 1mL/hr	15 X Weight (Kg)	Dopamine Amiodarone Dobutamine	5 - 20mcg/kg/min 5 - 15mcg/kg/min 2 - 20mcg/kg/min
10 mcg/kg/min = 1 mL/hr	30 X Weight (Kg)	Dopamine Amiodarone Dobutamine	5 - 20mcg/kg/min 5 - 15mcg/kg/min 2 - 20mcg/kg/min
20 mcg/kg/min = 1mL/hr	60 X Weight (Kg)	Lidocaine	20 - 50mcg/kg/min
25 mcg/kg/min = 1mL/hr	75 X Weight (Kg)	Esmolol	100 - 300mcg/kg/min
50 mcg/kg/min = 1mL/hr	150 X Weight (Kg)	Esmolol	100 - 300mcg/kg/min
Mcg/kg/hr	**DOSE in MG in a 50mL Syringe**		**Mcg/kg/hr**
10 mcg/kg/hr = 1mL/hr	0.5 X Weight (Kg)	Morphine	10 - 40mcg/kg/hr
20 mcg/kg/hr = 1mL/hr	1 X Weight (Kg)	Morphine	10 - 40mcg/kg/hr *Titrate up as required
Units/kg/hr	**DOSE in UNITS in a 50mL Syringe**	**Units/kg/hr**	
10 Units/kg/hr = 1mL/hr	10 X 50 X Weight (Kg)	Heparin – standard concentration	
Mg/kg/hr	**DOSE in MG in a 50mL Syringe**	**Mg/kg/hr**	
0.25 mg/kg/hr = 1mL/hr	12.5 X Weight (Kg)	Furosemide	0.25 - 0.5mg/kg/hr
Mg/kg/day	**DOSE in MG in a 50mL Syringe**	**Mg/kg/day**	
1 mg/kg/day = 1mL/hr	2 X Weight (Kg)	Phenoxybenzamine	0.5 - 2 mg/kg/day
Units/kg/min	**DOSE in UNITS in a 50mL Syringe**	**Units/kg/min**	
0.0001 Unit/kg/min = 1mL/hr	0.3 X Weight (Kg) (Max: 50 Units/50mL)	Vasopressin Sepsis: 0.0001 - 0.001units/kg/min Vasodilatory Shock post CV surgery: 0.001- 0.002 units/kg/min Brain death: 0.0003 units/kg/min Diabetes Insipidus: 0.0001 - 0.00025 units/kg/min	
Nanogram/kg/min	**DOSE in MICROGRAM in a 50mL Syringe**	**Nanogram/kg/min**	
5 nanogram/kg/min = 1mL/hr (0.005 mcg/kg/min = 1mL/hr)	15 X Weight (Kg)	Epoprostenol (Prostacyclin or Flolan)	5 - 40nanogram/kg/min
	DOSE in GRAM in a 40mL Syringe	**Mg/kg/hr**	
Standard concentration titrate as per order	1G in 40mL of sterile water = 25mg/mL	Thiopentone	2 - 4 mg/kg/hr

Refer to Paediatric Critical Care Medicine: Handbook of Clinical Practice. Department of Critical Care Medicine. Rev Nov 2001, Critical Care Unit, Hospital for Sick Children. Reviewed 2023

7.3 ACE Inhibitor Guideline

Aim

This guideline is designed for all medical and nursing staff involved in the management of children commencing on Angiotensin Converting Enzyme inhibitors (ACE inhibitors).

Indications

- Paediatric cardiology patients (both pre and post operative) with a significant left to right shunt that requires additional intervention above diuretic therapy.
- Severe aortic or mitral incompetence with left ventricular dilatation.
- Primary left ventricular dysfunction e.g. dilated cardiomyopathy or post chemotherapy left ventricular dysfunction.
- Primary systemic hypertension following thorough investigation of possible aetiology and in absence of renal disease – structural or functional.

Drug Information

Class: Angiotensin Converting enzyme inhibitor

Class		Drug Name	Half Life
I	(Active compound)	Captopril	1.9 hr
II	(Prodrug)	Enalapril	11 hr
III	(water soluble)	Lisinopril	12hr

ACE inhibitors inhibit the conversion of angiotensin I to angiotensin II. The primary site of action is the efferent arterioles of renal glomerulus with ACE inhibition causing vasodilatation and thus reducing afterload. There is also some cross reactivity on the bradykinin system as well as possible direct effects on cardiac reverse-remodelling.

Available preparations

Captopril: Licensed liquids available in 2 different strengths (Noyada®; 21 day expiry):
5mg/5ml to be used for doses <2.5mg
25mg/5ml to be used for doses ≥ 2.5mg

Tablets 12.5mg (Tablets can be crushed and dispersed in 5mls of water, 2.5mg/ml solution).

Enalapril: Tablets 2.5mg, 5mg and 10mg
Special 5mg/5ml liquid available

Lisinopril: Tablets 2.5mg, 5mg, 10mg and 20mg
Special 5mg/5ml liquid available

Side effects

The principle side effect is significant first dose hypotension; this can be more pronounced in the neonatal patient group and children on diuretic or other vasoactive medications. Renal dysfunction and hyperkalaemia, angioedema and dry persistent cough.

Contraindications

ACE inhibitors should not be used in patients with known hypersensitivity to ACE inhibitors or in patients with bilateral renovascular disease. Clear rationale to be documented if prescribing in these circumstances. If patient is intolerant to ACEi, consider use of hydralazine.

Commencing Captopril

Captopril should be commenced as an inpatient. The decision to commence captopril should be taken by the admitting consultant paediatric cardiologist or paediatrician with an interest under the direction of the consultant paediatric cardiologist.

Day case admission

This should be arranged by the consultant paediatric cardiologist via their secretary and the ward sister. This will be a full day admission and patients should ideally arrive by 09:00am on the day of admission.

Prior to admission, all patients should have an echocardiogram (Echo) performed within 6 weeks of admission. Renal function should be checked within one week of admission.

On arrival the patient will require:

- Medical clerking and nursing admission
  (Identify the Doctor responsible for prescribing the ACE inhibitor and the inpatient management)
- Baseline observations; Heart rate, Respiratory rate and Blood Pressure recorded, and weight measured. Identify appropriate physiological parameters with medical staff, and record on age appropriate observation chart. Calculate Paediatric Early Warning Score (PEWS) & escalate as appropriate
- Check Renal function and Echocardiogram have been performed as per instructions
- If patient is less than 6 months of age, consider siting a cannula when taking bloods in case there is a need for intravenous fluids due to hypotension.
- If all the above checks are satisfactory prescribe the medication.
- The test dose can be given prior to blood results being available unless there is reason to suspect renal dysfunction.
- Blood pressures should be monitored every 20 minutes.
- Note: some patients may require more frequent monitoring of BP; this should be assessed on an individual patient basis.
- It should be expected that all patients have a transient and limited fall in BP. However, if symptoms persist and treatment required, patient should be laid flat, legs elevated and a fluid bolus considered. Only a prolonged period of hypotension should be considered an adverse response.

Dosage:

CAPTOPRIL

……..mg 8 hourly PO

Test dose 0.05mg/kg to 0.1mg/kg PO
Increase as tolerated up to
0.5-1mg /kg tds. Monitor as per network
Guideline. **Max test dose 6.25mg.**
Max regular dose 25mg

Test dose: 0.1mg/kg (maximum test dose 6.25mg).

In neonates consider 0.05mg/kg test dose.

Providing the patient tolerates the test dose then a regular dose of 0.2mg/kg per dose three times a day should be prescribed. Depending on the preference of the admitting cardiologist this dose should be escalated in time to 0.5mg/kg per dose three times a day (higher doses of 1mg/kg dose may be indicated in selected patients). Up-titration of the dose should be done with caution in unstable or young patients, usually under inpatient supervision with careful monitoring of renal function/electrolytes.

Once patient has reached desired dose they should be discharged with a completed discharge letter with new dose prescribed for the G.P. Arrangements should be made for renal function to be checked within a week of discharge (at the patient's local hospital, locally or at their G.P. surgery; ensuring there is a system in place to check the results).

Further increases can be managed either as an outpatient or a subsequent admission. Monitoring of renal function is advised with every dose change.

If patients are on spironolactone and the captopril dose exceeds 0.5mg/kg then consider discontinuation of spironolactone.

Captopril test doses are not usually given at the same time as diuretics. However, prior to discharge it should be assessed, whether the patient can tolerate administration of the diuretics with the captopril at the same time, without causing significant hypotension.

[Note: for adolescent patients, please consider sensible test & discharge dosages with the tablet sizes available]

Conversion to Enalapril or Lisinopril

In older children a longer acting ACE inhibitor may be preferable. Conversion to either enalapril or lisinopril should only be undertaken when the patient has been proven to tolerate captopril trial. Drug labels should be used for all prescriptions.

Conversion relative to captopril dose

Enalapril	1mg enalapril = 7.5mg captopril	ENALAPRIL orally 1 month-11 years: initially 0.1mg/kg daily increased as required to 0.5mg/kg daily. 12-18 years: initially 2.5mg once daily. Maintenance 10-20mg daily for children. Max dose for 50kg and greater: 40mg daily. Can be given divided into twice daily dosing.
Lisinopril	1mg lisinopril = 5mg captopril	LISINOPRIL orally 12-18 years: initially 2.5mg once daily, increased in steps to maintenance dose of 10-20mg once daily. Maximum daily dose 35mg. Dose usually given once daily

Intolerance to ACE inhibitors

Some patients do not tolerate ACE inhibitors due to the persistent dry cough. A suitable alternative could be Losartan. Losartan is an angiotensin-II receptor antagonist that unlike ACE inhibitors does not inhibit the breakdown of bradykinin and other kinins and are thus less likely to cause the persistent dry cough.

There is no direct dose conversion from ACE inhibitors to Losartan.

Contributors: Network Medicines Group – guideline created 2019, reviewed 2023

7.4 Losartan guideline

Indications

- Large L→R shunt }
- Severe aortic regurgitation } but ACE inhibitor
- Severe mitral regurgitation } not tolerated
- Ventricular dysfunction }
- Hypertension (e.g. following coarctation repair) }
- Aortic enlargement (e.g. Marfan syndrome)

Side Effects

Important recognised side effects include:

- Hypotension
- Renal impairment
- Angioedema
- Tiredness

Dose

The initial dose is 0.5 mg/kg once daily, increasing to 1-2 mg/kg once daily according to response.

<50 kg, maximum dose is 50 mg once daily.

50 kg and over, maximum dose is 100 mg once daily.

If an ACE inhibitor is being substituted, a straight swap of losartan in place of the ACE inhibitor can be made without needing a test dose. Alternatives to losartan in older patients are valsartan or candesartan (licensed indications: hypertension and/or heart failure).

Irbesartan suspension is available as a special. Dose: up to 2 mg/kg given once daily. Doses of up to 4 mg/kg/day have been given in cases of Marfan syndrome with severe aortopathy.

Reviewed 2019 – with thanks to Neil Dawson

7.5 Amiodarone guideline

Mode of Action

An antiarrhythmic agent which prolongs the action potential duration in both the atrial and ventricular myocardium (class III antiarrhythmic)

Intravenous amiodarone:

Cautions

- Use great care in neonates and in patients with fractional shortening <25%.
- Consider concomitant use of IV inotrope (dopamine, adrenaline) if fractional shortening is <25% or patient has signs of shock
- Consider giving the loading dose over 60-240 minutes, particularly if the patient is haemodynamically unstable (think – *would DC shock be safer and more effective?*)
- Dilute according to manufacturer's recommendation (dilutions of <0.6 mg/mL, i.e. <600 µmL are not stable– see advice on mixing up the infusion below)
- Give into a central vein if possible – watch for phlebitis if given peripherally
- Watch for hypotension/bradycardia – if these are seen, reduce or stop the infusion
- Check baseline U&E, LFT and TFT – also, lung function in older children having elective rhythm treatment; weekly LFTs whilst on IV amiodarone
- Watch effect on QTc – accepted QTc of up to 470 msec
- Continuous ECG monitoring when starting IV treatment then a 12 lead ECG once the oral therapy has been continued for 3 days without iv amiodarone
- Advise use of high protection sun block if being used long-term – to continue for 6 months after stopping amiodarone; where possible avoid phototherapy lights for treatment of neonatal jaundice due to risk of blue-grey syndrome.

Loading dose

Stable patient, non-neonate, and FS≥25%
5 mg/kg infusion over 30 minutes

Alternatively, if the patient is unstable, is a neonate or if the FS is <25%
Either
5 mg/kg loading dose over 60-240 minutes
or
25 microgram/kg/min for (1.5 mg/kg/hr) 4 hours
Consider concurrent use of dopamine or adrenaline

Maintenance infusion

5-15 microgram/kg/min (300-900microgram/kg/hour) adjusted according to response, (max. 1.2grams in 24hours)

Preferably administer via a **central line**. If central venous access is not available, use a **large peripheral vein** and monitor the injection site closely. Concentrations exceeding amiodarone 2mg in 1mL should always be given via a central venous access device except in extreme clinical emergency.

Mixing up the infusion

Amiodarone is not compatible with sodium chloride solution – use 5% glucose

EITHER:
Take 1 mL of 50 mg/mL solution and dilute to 50 mL with glucose 5% to give a 1 mg/mL solution.
0.3 mL/kg/hr = 300 micrograms/kg/hour = 5 microgram/kg/minute.

OR – if fluid restricted:

Take 3 mL of 50 mg/mL solution and dilute to 25 mL with glucose 5% to give a 6 mg/mL solution (maximum stable concentration).

0.05 mL/kg/hr = 300 micrograms/kg/hr = 5 microgram/kg/minute.

Compatiblities
Adrenaline, atracurium, calcium gluconate, dobutamine, dopamine, esmolol, fentanyl, insulin,labetalol, midazolam, milirione, morphine, noradrenaline, phenylephrine, vancomycin, vasopressin, vecuronium. **These drugs should NOT be diluted with sodium chloride 0.9%.**

Incompatiblities
Aminophyllin, furosemide, heparin, potassium acid phosphate, phosphate infusions, sodium bicarbonate, sodium chloride, sodium nitroprusside. If there is no compatibility information for specific drugs, do not assume compatibility, use a separate line.

Expiry
Syringe must be changed every 24 hours

Older teenagers and adults with VT
150mg IV in 10 min for VT
Then 1mg/min for 6 hours
Then 0.5 mg/min maintenance until control of rhythm

Oral amiodarone:
Neonates
Refer to BNFc

Under 12s (excluding neonates)
Load: 5 mg/kg/dose given three times daily for 7 days
then 5 mg/kg/dose given twice daily for 7 days
then 5 mg/kg/day given once daily as maintenance

Over 12s
Load: 200 mg three times daily for 1 week
then 200 mg twice daily for 1 week
then 200 mg once daily for 1 week

The correct maintenance dose is the minimum dose that successfully controls the rhythm.

Tablet strength available 100mg & 200mg; oral liquid can be prepared specially. Amiodarone tablets can be crushed and dispersed in water.

Drug interactions and long-term monitoring
Amiodarone-Digoxin – reduce digoxin dose by at least 30%, check levels.

Amiodarone-Flecainide – may take two weeks or more to develop fully. Reduce the flecainide dose by up to 50% and monitor for flecainide adverse effects.

Amiodarone-Warfarin – enhanced effect, so monitor INR carefully on starting treatment and changing doses.

Check follow-up LFT and TFT 6 monthly. Watch lung function long term. Corneal microdeposits may occur, but these reverse on stopping treatment. Photosensitivity (sunburn) of exposed areas occur frequently. Sunblock creams are required.

Counselling

Ensure the parents have received an information leaflet before discharge. These can be found at https://www.medicinesforchildren.org.uk/amiodarone-abnormal-heart-rhythms-0.

Contributors: Orhan Uzun, Clara Danielson, Dirk Wilson – Reviewed 2023

7.6 Aspirin in Cardiac Disease

In general, use of aspirin in children <16 years is not recommended because of the risk of Reye's syndrome. If aspirin is to be given long-term, ensure children >6 months of age are vaccinated against influenza and consider use of varicella vaccination.

The indications for the use of aspirin are set out in Section 6.1. The table below summarises the different doses used for different indications:

Indication	Dose	Comment
Anti-platelet action, e.g. post cardiac surgery, primary prevention of thrombosis	3-5 mg/kg given once daily	Use 5 mg/kg/dose in neonates
Kawasaki disease	See section 6.13	Meta-analysis comparing this dose (30-50 mg/kg/day) with the AHA/Japanese recommendation (80-100 mg/kg/day) found no difference in outcomes. Switch to lower dose either at 2 weeks, or once inflammation has settled.
Acute phase	7.5 – 12.5 mg/kg/dose given 4x daily (neonate 8 mg/kg/dose 4x daily)	
Convalescent phase	3-5 mg/kg/dose given once daily, continue for minimum of 6 weeks	
Pericarditis	12.5 – 20 mg/kg/dose given 4x daily	Continue until acute inflammation settles fully
Rheumatic fever	Either: 25 mg/kg/dose 4x daily tapering to 15 mg/kg/dose given 4x daily	Give for 2-3 weeks, then taper to lower dose once symptoms resolve
	Or 12.5-15 mg/kg/dose given 4x daily	Continue for up to 12 weeks

7.7 Carvedilol guideline

Aim This guideline is designed for use by all medical and nursing staff involved in the care of paediatric cardiology patients commenced on carvedilol either as part of on-going inpatient management or as a day case admission.

Indications

Carvedilol is a non-selective beta-adrenoceptor and alpha-1 adrenergic antagonist causing vasodilatation. In adult practice it is licensed for use in hypertension and chronic heart failure. It remains unlicensed in the paediatric population however several studies have demonstrated

an improvement in symptoms and cardiac function in children with severe cardiac left ventricular systolic dysfunction and dilated cardiomyopathy.

Drug Information

Preparations
Tablets available in 3.125mg, 6.25mg, 12.5mg and 25mg.

Side effects
The principle adverse effect is hypotension. Possible bradycardia in higher doses reported. Other reported effects are hyperglycaemia, gastrointestinal disturbance (preparations may contain a small amount of lactulose) and rash.

Contra-indications
Relative contraindications are a history of asthma/reactive airways disease. Rhythm disturbance with sinus bradycardia and/or second degree or third degree heart block. Hepatic impairment.

Commencing Carvedilol
The decision to commence carvedilol can only be taken by a consultant Paediatric Cardiologist and may be coordinated with the paediatrician with an interest if the patient is determined stable. The responsible consultant should be identified prior to the admission.

Carvedilol is usually considered after full medical treatment with an angiotensin converting enzyme inhibitor (ACEi), when the cardiac failure is considered stable. This is usually at least 2-4 weeks after the discontinuation of any inotropic support.

Daycase admission
On admission and prior to the first dose the patient should have:

- Medical clerking and nursing admission
- Baseline observations performed (Heart rate, resp rate and blood pressure) and weight measured
- Bloods taken for baseline renal and hepatic function (U & E's and LFTs)
- Baseline ECG (for Heart block and bradycardia) and Echocardiogram performed (for Function; LV dimensions and Fractional shortening)
- Have IV access
- Prescribe the test dose

Test dose of carvedilol 0.05mg/kg can be prescribed. Max test dose 3.125mg.

- Blood pressure should be monitored every 30 minutes for 2 hours post administration.
- If the patient tolerates the test dose then this dose should be prescribed as a twice daily regime.
- The timing of the dose should be spaced initially, as not to coincide with other vasoactive drugs (captopril, diuretics etc). We recommend 1 hour spacing between vasoactive medications for the test dose. Consideration of the timing of the medications should be made prior to discharge.
- They should have a discharge summary generated to take with the patient and a copy sent to the G.P.

Subsequent increases in dose
Patient can have their dose increased at 4 weekly intervals in the outpatient setting if stable. At each review an echocardiogram should be performed, looking specifically at function and

baseline observations recorded.The right arm BP should be taken and clearly recorded in the notes and a 12 Lead ECG performed. Dosing guidance is set out below:

Slow titrating of dosage in Outpatient Setting

	Initial	Week 4	Week 8	Week 12
Dose	0.05mg/kg BD	0.1mg/kg BD	0.2mg/kg BD	0.3mg/kg BD
Maximum Dose	3.125mg BD	6.25mg BD	12.5mg BD	25mg BD

Doses may be increased every 4 weeks to a maximum of 0.35mg/kg/dose (25mg) BD **or** until side effects limit further dose increases e.g. symptoms of worsening CHF.

Initial dose to be given as an inpatient with subsequent dose increases as an out- patient If patient is stable.

Quick titrating of dosage in stable heart failure- Inpatient only at the cardiac centre

- In some exceptional circumstances an accelerated regime can be used to increase the dose in some stable patients who are in hospital
- This should be avoided in hospitals outside the cardiac units in Cardiff and Bristol Childrens' Hospital

	Initial (Day 1)	Day 3	Day 5	Day 7	Day 9
Dose	0.025mg/kg BD	0.05mg/kg BD	0.1mg/kg BD	0.2mg/kg BD	0.3mg/kg BD
Max dose	1.5mg BD	3.125mg BD	6.25mg BD	12.5mg BD	25mg BD

Doses may be increased every 2 days if tolerated to a maximum of 0.35 mg/kg (25mg) BD **or** until side effects limit further dose increases e.g. symptoms of worsening CHF.

NOTE: If symptoms of worsening CHF occur stop the carvedilol.

Following the final increase to a maximum of 0.7mg/kg/day then further follow up should be in the outpatient clinic. Please inform the patient's cardiologist of admission and ensure an out-patient appointment has been arranged. Blood test results will need to be checked by the ward medical staff/Registrar that admitted the patient.

Dose interactions

Patients on Digoxin should have their dose reduced by approximately 25% if prescribing carvedilol concomitantly.

RELATED DOCUMENTS

Carvedilol as therapy in pediatric heart failure: an initial multicenter experience. Bruns L *et al.* J Pediatr. 2001 Apr;138(4):505-11.

Carvedilol increases the systemic bioavailability of oral digoxin. De Mey C, Brendel E, Enterling D. Br J Clin Pharmacol (1990) 29 , 486-90.

Digoxin-Carvedilol interaction in children. Ratnapalan S, Griffiths K, Costei AM, Benson L, Koren G. J Pediatrics (2003) 142, 572-4.

Medicines for Children Information for Parents and Carers. Carvedilol for Heart Failure.
BNFC2017-2018
Guy's & St Thomas' Paediatric Formulary 9th Edition
Leeds Paediatric Formulary

Contributors: Network Medicines Group – guideline created 2019

7.8 Entresto in children

Entresto is a drug combination of the BNP receptor antagonist sacubutril and and the angiotensin receptor blocker valsartan. It may be used in children with severe LV dysfunction. It has been licensed for use by the FDA but is not licensed in children in the UK. If its use is being considered, refer to:

label (fda.gov)

For review 2026

7.9 Flecainide for SVT

Disclaimer:

This has been produced as a guide; each individual patient should be considered whether treatment is clinically appropriate. If needed, please discuss further with specialist teams.

Flecainide should only be used under the direct supervision of a paediatric/adult cardiologist

Indication

Flecainide is a safe and effective treatment for supraventricular tachycardia but should be used with caution in the setting of congenital heart disease and infancy. It should be avoided in patients with significantly impaired left ventricular function (LV).

It is an anti-arrhythmic agent used in the treatment of: resistant re-entry supraventricular tachycardia, ventricular tachycardia, ventricular ectopic beats, arrhythmias associated with accessory conducting pathways e.g. Wolff-Parkinson-White syndrome, and paroxysmal atrial fibrillation. Flecainide can also be used to control fetal supraventricular arrhythmias, as it crosses the placenta.

In the treatment of atrial fibrillation and flutter it must be combined with an AV node blocking agent such as propranolol, atenolol or digoxin.

Presentation

Tablets: 50mg, 100mg
Modified release capsules:200mg
Liquid: 25mg in 5ml (Specials supplier)
Injection:10mg/ml x 15ml ampoule

Class of drug

Local anaesthetic type anti-arrhythmic compound (Class Ic anti-arrhythmic)

Method of action

Flecainide reduces the maximum rate of depolarisation in heart muscle and thereby slows conduction particularly in the His-Purkinje system. It has a profound effect on conduction in accessory pathways, especially on retrograde conduction, and markedly suppresses ventricular ectopic foci. It is a local anaesthetic agent.
Note: has a negative inotropic effect and can itself precipitate serious arrhythmias.

Initial dosage (adjustment according to plasma-flecainide level)

FLECAINIDE
……..mg…….hourly PO

Neonate-12 years: 1-2mg/kg 8-12 hourly
12-18 years: 50-100mg 12 hourly
See network guideline.
Monitor levels.
Separate doses from milk feeds by 30 minutes.

Neonate 2 mg/kg/dose given 2-3 three times daily (caution if total > 8mg/kg/day) – the Cardiff unit would advise 1-2 mg/kg/dose bd dosage in the first instance – assess clinical response, ECG and drug levels
1 month – 12yrs 2mg/kg/dose given 2-3 three times daily (maximum 8mg/kg/day)
12 – 18 yrs 50 -100 mg orally twice daily (maximum 300mg/day. In heavier children with VT, doses of up to 400mg/day may be considered)

Oral administration

- Caution; milk, infant formula, and dairy products may interfere with the absorption of flecainide.
- The liquid has a local anaesthetic effect and should be given at least 30 minutes before or after food. **Do not store the liquid in refrigerator as precipitation occurs**.
- Caution is needed when patients are weaning from milk feeds onto solids.
- We would not recommend IV flecainide as replacement for Oral in NBM patients

Note: if the patient is NBM or acutely unwell (e.g. diarrhoea and vomiting), in particular an infant, who is mainly on milk feeds, consideration must be made that flecainide toxicity can occur and there should be a discussion with the consultant paediatric cardiologist on service, as to whether treatment is reduced or omitted.

Pharmacokinetics

Flecainide is rapidly and almost completely absorbed from the gastrointestinal tract following oral administration. Oral bioavailability is 90-100%. Flecainide is metabolised extensively in the liver to two major metabolites (meta-0-dealylated flecainide and its lactam). Both metabolites undergo extensive conjugation and are not considred to be pharmacologically active.
Flecainide and its metabolites are excreted largely in the urine with 25% of the dose unchanged drug. Plasma protein binding is approximately 40%.

Renal Impairment

Reduce dose (depending on degree of impairment) and monitor plasma levels.
Refer to BNF-C for dosing recommendations.

Hepatic Impairment

Avoid (unless benefit clearly outweighs risk), or reduce done in severe impairment
Monitor plasma levels

Initial Monitoring

Baseline ECG:
Take trough sample after 2-4 days, immediately prior to next dose*.
Therapeutic range: 200-800micrograms/litre (Cardiff Lab normal range 0.15 – 0.9 mg/L)
Approx time to steady state: 2 days
Repeat ECG at the same time, if QRS duration has increased by >25 %, reduce the dose.

Usual turnaround is 7-10 days unless the sample is marked “urgent”.

*In Cardiff we have typically taken samples 6 hours post-dose and have not seen problems with drug levels.

Long Term Monitoring

In children who have a total daily dose of > 6mg/kg/day should have regular ECG and drug levels 6 monthly.

Interactions

For full list of interactions, see Appendix 1of BNF-C. Some relevant cardiac interactions are:

- Plasma concentration of flecainide increased by amiodarone (suggest using half dose of flecainide)
- Increased risk of myocardial depression and bradycardia when flecainide given with beta-blockers
- There is conflicting evidence about the interaction of flecainide and digoxin. Some suggest an increase of 15 -25% in the digoxin levels. Consider digoxin level monitoring on the administration of flecainide.

Contra-indications

Heart failure
Impaired LV function
Haemodynamically-signficant heart valve disease
Sinus node dysfunction
Atrial conduction defects
Second degree AV block, bundle branch block or distal block (unless rescue pacing available)

Precautions

- Correct electrolyte imbalances- increased cardiac toxicity with flecainide when hypokalaemia occurs.
- Flecainide is known to increase endocardial pacing thresholds – i.e. increase the amount of energy needed to effectively pace the myocardium. This effect is reversible on stopping the medication. Flecainide should thus be used with caution in all patients with permanent pacemakers or temporary pacing electrodes, and should not be administered to patients with existing poor pacing thresholds or non-programmable pacemakers, unless suitable backup pacing is available.
- The IV route should only be used with continues ECG monitoring and where full cardiopulmonary resuscitation can be readily available (HDU/PICU).

Side effects

There are many listed side effects. The most common include:
Pro-arrhythmic effects
Dyspnoea,
Dizziness
Asthenia
Fatigue
Fever
Visual disturbances
Oedema.
For a full list of side effects
reported, see the drug data sheet and MHRA (Medicines and healthcare products regulatory agency) data analysis prints. [1,2]

Toxicity
Even therapeutic doses may induce or aggravate arrhythmias. In cases of overdose, cardiac features predominate with delayed conduction through the myocardium and depression of myocardial contractility. Bradycardia and tachycardia have been reported. Other features include ECG abnormalities, hypoxia, metabolic acidosis, hypotension and convulsions. No specific antidote is known. Contact Toxbase on 0344 892 0111 if you are concerned about flecainide toxicity in a patient.

Pregnancy
Used to treat maternal and fetal arrhythmias under specialist care.

Breast feeding
Flecainide is excreted in human milk and appears in concentrations which reflect those in maternal blood, but is not known to be harmful.

References

[1] Summary of Product Characteristics. Flecainide.
http:www.medicines.org.uk/emc/medicine/27678
[2]MHRA
http://www.mhra.gov.uk/drug-analysi-prints/
[3] British National Formulary for Children(BNF-C) 2017-2018
[4] Medicines for Children 2003 RCPCH
[5] Guys and St Thomas' Hospital Paediatric Formulary 9th Edition
Contributors Network Medicines Group – guideline created 2019

7.10 Hydralazine for Treatment of Heart Failure
The recommended dosing schedule is shown below:

- Day 1 0.25 mg/kg/dose given twice daily
- Day 2 If tolerated increase to 0.25 mg/kg/dose three times daily
- Day 3 If tolerated increase to 0.5 mg/kg/dose three times daily
- Day 5 onwards, depending on response – increase to 1 mg/kg/dose three times daily; doses of up to 2 mg/kg/dose can be used

Look out for side effects (tachycardia, flushing, hypotension, GI disturbance and, if used long-term, lupus-like syndrome – stop if skin rash develops). Monitor FBC.
Contributors: Neil Dawson, Orhan Uzun, Dirk Wilson – guideline reviewed 2023

7.11 Ivabradine use in Children

Aim
This guideline is designed for all medical and nursing staff involved in the management of children commencing on Ivabradine.

Indications
Ivabradine is licensed in the UK for use in adults for the treatment of angina (for patients in sinus rhythm) and heart failure.
It is not licensed for use in children, so any prescription in under 18s is "off license" and the family should be aware of this.

Indications for use in children include:
Postural orthostatic tachycardia syndrome (POTS) in older children and adolescents

Inappropriate sinus tachycardia leading to symptoms and where other treatable causes have been ruled out
Atrial tachycardia
Congenital junctional ectopic tachycardia
Heart failure as an adjunct treatment, e.g. if carvedilol has not been tolerated

Contraindications

- Acute myocardial injury
- Cardiogenic shock
- Congenital QT syndrome
- Do not initiate
    - for chronic heart failure if heart rate below 75 beats per minute
    - immediately after cerebrovascular accident
    - patients dependent on pacemaker
    - second- and third-degree heart block
    - severe hypotension
    - sick-sinus syndrome
    - sino-atrial block
    - unstable angina
    - unstable or acute heart failure

Drug Information

Ivabradine acts by reducing the heart rate via specific inhibition of the pacemaker current (funny current, I_f) – the action is most marked for cells with a steep depolarising potential (2.g. sino-atrial node tissue).

Preparations

Tablets are available as 5mg and 7.5mg.

At present no commercially made solution is avaialble, therefore, for doses of <2.5 mg, 5mg tablets may be crushed and diluted in 10ml of water to give a 1mg/2ml solution.
For non adherent children the tablets can be crushed and mixed with yogurt or soft food for administration.

Dosage and Commencement of Treatment

Before starting, check ECG and echo.
Ensure normal, AV node function (no AV block), QT interval and normal (or stable) systemic ventricular function.

<u>Heart rhythm indications or POTS</u>
Children <1 year of age.
Starting dose 0.02 mg/kg/dose given twice daily,
After 1-2 weeks repeat ECG; if satisfactory increase to 0.05 mg/kg/dose twice daily
After 1-2 weeks repeat ECG; if satisfactory increase to 0.1 mg/kg/dose twice daily
Discuss with consultant if doses of 0.15mg/kg/dose or 0.2mg/kg/dose required.

Weight of <40 kg and >1year of age
Starting dose 0.05 mg/kg/dose given twice daily
After 1-2 weeks repeat ECG; if satisfactory increase to 0.1 mg/kg/dose twice daily
After 1-2 weeks repeat ECG; if satisfactory increase to 0.15 mg/kg/dose twice daily
After 1-2 weeks repeat ECG; if satisfactory increase to 0.2 mg/kg/dose twice daily
Maximum dose 7.5 mg twice daily

Weight of >40 kg
Starting dose 2.5 mg twice daily
After 1-2 weeks repeat ECG; if satisfactory increase to 5 mg twice daily
Maximum dose 7.5 mg twice daily

Heart failure indications
Doses as above, but 2-4 weeks between increments – assess ventricular function on a regular basis
The aim is to see a more than 20% reduction from baseline heart rate without inducing symptoms of bradycardia.
Lowest acceptable heart rate compared to age:

Age	Lowest acceptable heart rate on treatment
6-12 months	>80 bpm
>1 year and <3 years	>70 bpm
3-18 years	>50 bpm

Interactions
See adult BNF for a comprehensive list.
Caution with (or avoid)

- Amiodarone - Ivabradine is predicted to increase the risk of torsade de pointes when given with amiodarone.
- Beta-blockers – increase risk of bradycardia and AV block
- Digoxin – Both ivabradine and digoxin can increase the risk of bradycardia
- Flecainide – Both ivabradine and digoxin can increase the risk of bradycardia
- Grapefruit juice – predicted to increase the exposure to ivabradine; manufacturer advises avoid
- Macrolide antibiotics – increased risk of torsades des pointes
- Rifampicin – predicted to decrease the exposure to ivabradine. Manufacturer advises adjust dose.
- Sildenafil - Ivabradine is predicted to increase the risk of torsade de pointes when given with sildenafil. Manufacturer advises avoid.
- Sotalol – both ivabradine and sotalol can increase the risk of bradycardia.

Side Effects
Common or very common:
Arrhythmias such as bradycardia and atrioventricular block
Dizziness; headache (first month of treatment); hypertension; vision disorders (luminous phenomena (phosphenes))

Uncommon
Abdominal pain; angioedema; constipation; diarrhoea; eosinophilia; hyperuricaemia; hypotension; muscle cramps; nausea; QT interval prolongation; skin reactions; syncope; vertigo

Monitoring
Regular ECG assessment is advised. If the indication is heart failure, regular echo assessment is advised.

Discontinuation
Stop ivabradine if there are concerning side effects, including symptomatic bradycardia.

There is no need to reduce the dose gradually in this scenario.
Where the indication is heart failure, if ivabradine is being discontinued because function has recovered, it would be sensible to reduce the dose over the space of 2-4 weeks and assess response.

References

Bonnet D, Berge F, Jokinen E et.al. (2017) 'Ivabradine in Children With Dilated Cardiomyopathy and Symptomatic Chronic Heart Failure', *Journal Of The American College Of Cardiology,* 70(10), pp. 1262-1272.
Dieks J,K, Klehs S, Muller M,J. et.al (2016) 'Adjunctive ivabradine in combination with amiodarone: A novel therapy for pediatric congenital junctional ectopic tachycardia.', *Heart Rhythm,* 13, pp. 1297–1302.
Summary of Product Characteristics – Ivabradine 5mg Film coated Tablets. Accord-UK Ltd. Accessed via https://www.medicines.org.uk/emc/product/9084/smpc on 24/06/19 [date of revision of the text 20/03/2019].

Contributors: Neil Dawson, Dirk Wilson – Guideline reviewed 2023

7.12 Magnesium Sulfate for Arrhythmia or Pulmonary Hypertension

Treatment of PPHN – refer to BNFc.

Treatment of torsades de pointes in a child 1 month to 18 years:
Use magnesium sulphate heptahydrate, 25-50 mg/kg (equivalent to 0.1 to 0.2 mmol/kg) given over 10-15 minutes.
Maximum dose = 2 g (equivalent to 8 mmol). Repeat dose once if necessary.

Reference: BNFc – Reviewed 2023

7.13 Midodrine use in children

Aim

This guideline is designed for all medical and nursing staff involved in the management of children commencing on Midodrine.

Indications

Midodrine is licensed in the UK for use in adults for the treatment of severe postural orthostatic hypotension due to autonomic dysfunction (for example, primary autonomic failure; Parkinson's disease/ multiple system atrophy; diabetic autonomic neuropathy; postural orthostatic tachycardia syndrome when corrective factors have been ruled out and other forms of treatment are inadequate).

It is not licensed in children and can only be prescribed on a named patient basis through the hospital pharmacy.

It is ordinarily used in older children/adolescents aged 12+ in the treatment of vasovagal syncope or POTS where this has been established with a head-up tilt test or where the history is clear and symptoms are severe.

Contraindications

Raynauld's phenomenon and other conditions where there is blood vessel constriction
Heart conduction disturbance
Heart failure
Hypertension
Hyperthyroidism
Urinary retention
Phaeochromocytoma

Drug Information

Midodrine is a pro-drug which is converted to desglymidodrine and stimulates peripheral alpha adrenergic receptors causing vasoconstriction of the venous system and increased peripheral arterial resistance, resulting in an increase in BP. Midodrine reaches peak concentration in the blood about an hour after swallowing a tablet, but the effect is brief, with levels falling to half about 2-3 hours later.

Dosage

Age range 12-18 years old
Initial dose: 2.5mg 2-3 times daily.
Increase if necessary in 2.5mg increments weekly, until an optimal response is obtained, up to a maximum dose of 10mg three times a day. Midodrine can be taken with or without food.

Midodrine is available as 2.5mg and 5mg tablets. Tablets can be taken whole or crushed and mixed with water for administration.

Doses should be taken during the **active part of the day** to avoid supine hypertension.

Midodrine should not be taken within four hours of expected recumbency, e.g. avoid giving after 6 pm.

If symptoms of postural hypotension recur - check compliance and that non-pharmacological strategies are being appropriately applied – the family should report this to the cardiac nurse specialist.

Patients will be informed to contact the cardiac nurse specialist or GP immediately if any of the following symptoms of supine hypertension (systolic hypertension > 160mm Hg) occur: chest pain, palpitations, shortness of breath, headache and blurred vision.

Interactions

See adult BNF
Digoxin risk of heart block
Beta-blockers - risk of bradycardia
Fludrocortisone – risk of hypertension – watch BP carefully if used ocncurrently
Antihistamines
Thyroid hormones
Over-the-counter decongestant remedies often contain related compounds, pseudoephedrine and phenylephrine

Side Effects

Most commonly reported-
Piloerection (goosebumps), pruritus and/or paraesthesia of the scalp, dysuria.

Commonly reported-
Paraesthesia, pruritus, headache, supine hypertension (dose dependent effect), nausea, dyspepsia, stomatitis, chills, flushing and rash, urinary retention.

Uncommon
Sleep disorders, insomnia, restlessness, excitability, irritability, reflex bradycardia, urinary urgency.

Rare

*T*achycardia, palpitations, abnormal hepatic function and raised liver enzymes.

Frequency not known – anxiety, confusional state, abdominal pain, vomiting, diarrhoea.

Monitoring

U&E/creatinine/LFT; lying and standing BP
Review of response to treatment; the frequency of which depends on patient.
Supine BP at three months and then six monthly thereafter; consider checking more often if Midodrine is used in combination with Fludrocortisone (consider 24 hour BP).
If any concerns check U&E/creatinine/LFT.

Discontinuation

If treatment is being discontinued due to side effects, it can stopped immediately.
If treatment is being discontinued because symptoms have improved, it would be sensible to reduce the dose slowly over 2-4 weeks in case symptoms recur.

Reference: Pediatr Pharamotherapy 2014;20(4) *http://www.medicine.virginia.edu/clinical/departments/pediatrics/education/pharm-news/home.html*

Contributors: Neil Dawson, Dirk Wilson, C&V Midodrine Guideline
Pharmacy & Medicines Management Intranet - Midodrine prescribing guidelines.pdf - All Documents (sharepoint.com)
Guideline reviewed 2023

7.14 Propranolol for the Treatment of Capillary Haemangiomas

Infantile Haemangiomas (IH) are the most comon benign vascular tumours of infancy. While the mechanism of action on IH is still under investigation propranolol has been shown to to induce a better and faster response in IH management than systemic steroids, with fewer side effects.

Cardiology will only be involved if there is a clinical cardiovascular concern by the prescribing team.

Refer to the C&V guidleline:
General Paediatrics Clinical Portal - infantile-haemangiomas.pdf - All Documents (sharepoint.com)

7.15 PGE infusion protocol (NB risk of apnoea)

Indications for prostaglandin E therapy:

Physiology	Diagnosis	Presentation
Low pulmonary blood flow	Critical PS Pulmonary atresia Tricuspid atresia	Profound cyanosis ± Murmur (Cardiogenic shock)
Low systemic blood flow	Severe CoA HLHS Interrupted aortic arch	Cardiac failure Cardiogenic shock Poor or unequal pulses
Inadequate mixing	Transposition of the great arteries Obstructed TAPVD*	Cyanosis Tachypnoea Collapse

*If obstructed TAPVD is confirmed, PGE infusion is not recommended.

Prescribing
Prostaglandin E must be prescribed by brand name to prevent risk of errors. Products available are PGE-2 (dinoprostone- 1st line) and PGE-1 (alprostadil – 2nd line). PGE-1 is the licensed preparation, but is more costly. The two forms are equally effective in keeping the ductus open. If side effects, such as fever or shivering, are intolerable with one formulation, swap to the other form.

Administration and Dosage
Prostaglandin E can be given peripherally or centrally, with either glucose 5% (preferred) or sodium chloride 0.9%.

Wt x 30 = number of **microgram** of PGE added to a **total volume** of 50 mL of 5% or 10% dextrose.
With this dilution, 1 mL/hr gives a dose of 10 nanogram/kg/min

Starting dose is 5-10 nanogram/kg/min
In unwell patients doses of up to 50 nanogram/kg/min can be used, and titrated downwards depending on response. There is an association between the use of high-dose PGE and necrotising enterocolitis.

Side effects
The common side effects are apnoea, hypotension, fever, tachycardia, shivering and looking flushed.

The major (and most common) complication on starting therapy is apnoea requiring ventilator support.
Apnoea is a less likely a complication on a dose of <15 nanograms/kg/min.
Apnoea as a side effect normally occurs within 1 hour after starting prostaglandin E, or if the dose is increased.

Reviewed 2023 – with thanks to Neil Dawson, Clara Danielson and Susie Gage

7.15 Rivaroxaban use in cardiac patients

Aim
This guidance supports the use of Rivaroxaban in children with underlying heart conditions.

Indications
The drug is licensed in term neonates and children for the treatment and prophylaxis of venous thromboembolism.
This intention is to use Rivaroban off-license in selected cardiac patients as an alternative to warfarin **in children ≥12 kg** with:

- Dilated cardiomyopathy (DCM) deemed to be at risk of thromboembolism due to poor cardiac function
- Fontan patients

Rivaroxaban can also be used in keeping with its product license in any aged child with venous thromboembolism – refer to BNFc for prescribing advice.
It should NOT be used in patients with prosthetic heart valves or in patients with anti-phospholipid syndrome.

When initiating patients on to rivaroxaban for a cardiac indication please make parents/guardians aware that the medication is currently not licensed for this use.

Absolute contraindications

- Patients with hepatic disease associated with coagulopathy and clinically relevant bleeding risk, including cirrhotic patients
- Patients with active clinically significant bleeding
- Hypersensitivity to the active substance or to any of the excipients
- Concomitant treatment with any other anticoagulants, e.g. unfractionated heparin (UFH), low molecular weight heparins (e.g. enoxaparin, dalteparin) heparin derivatives (e.g. fondaparinux.), oral anticoagulants (warfarin, dabigatran, apixaban) except under specific circumstances of switching anticoagulant therapy
- Other haemorrhagic risk factors, as with other antithrombotics, rivaroxaban is not recommended in patients with an increased bleeding risk such as:
    - congenital or acquired bleeding disorders
    - uncontrolled arterial hypertension
    - vascular retinopathy
    - bronchiectasis or history of pulmonary bleeding
- **Metallic heart valve** requiring anticoagulation (warfarin preferable)

Relative contraindications

- Renal impairment
- Underlying lesion coveying significant risk for major bleeding (e.g. current or recent GI ulceration, recent brain or spinal bleeding, injury or surgery, jejunal bleeding, oesophageal varices, vascular malformations (especially intraspinal or intracerebral)

Drug information

Rivaroxaban (Xarelto^{TM}) is one of the direct oral anticoagulant (DOAC) drugs. It is a direct factor Xa inhibitor, targeting free and bound Factor Xa in the prothrombinase complex. Onset of action is within 2-4 hours and the half-life in young infants is 1.6 hours and in adolescents is 4.2 hours.
Rivaroxaban bioavailability is reduced if taken on an empty stomach. It should therefore always be given WITH FOOD.

Tablets can be crushed and administered via NG tube or PEG, however as absorption is site dependent administration via naso-jejunal or surgically placed jejunostomy tubes is NOT recommended as it will lead to reduced adsorption

Drug interactions

See BNFc for full list. Cardiac drug interactions include:

- Amiodarone (increased effect of Rivaroxaban, but dose reduction is not advised)
- Other antiplatelet or antithrombotic agents
- Epoprostenol and Treprostinil
- Important non-cardiac inducers and inhibitors of CYP3A4, e.g. azoles, phenytoin

Side effects

See BNFc for a full list. The most important side effect is risk of haemorrhage.

Available preparations

Rivaroxaban is available as granules for reconstitution as an oral suspension (1 mg/mL), but the shelf life is very short. It is also available in tablet form (2.5 mg, 10 mg, 15 mg, 20 mg).

Note: A 100ml bottle after reconstitution only contains 51.7ml of liquid
The 250ml bottle after reconstitution contains 103.4ml.

Commencing Rivaroxaban

Use table below for doses in children 12 kg and above.

Please see the separate Eligibility Criteria Table (Rivaroxaban Appendix 1) to be completed for all new patients.

Patients and parents/guardians must be counselled on the new medication. Please print the Counselling Checklist (see Rivaroxaban Appendix 2) and add a signed copy to the patient notes.

Newly Initiated Patients: Heparin or enoxaparin must be administered for a minimum of five days prior to starting rivaroxaban. On day 6 start rivaroxaban when the next dose of enoxaparin is due or when unfractionated heparin is stopped.

Switching from warfarin to rivaroxaban: start rivaroxaban when INR ≤ 2.5. Once Rivaroxaban has been given, the INR will not be an accurate measure of anticoagulant activity.

Dosage

Please see BNFc for dosing if patient is <12kg.

For patients 12 kg and above:

Weight	Dose	Frequency
12 – 29.9 kg	5 mg	Twice daily
30 – 49.9 kg	15 mg	Once daily
50 kg +	20 mg	Once daily

Note: Use with caution in obesity. Discuss with haematology and consider rivaroxaban levels if patient is >120kg or above the 97th percentile for weight.

For children under 12 kg, refer to BNFc for dosage.

Monitoring requirements

Patients should be monitored for signs of bleeding or anaemia; treatment should be stopped if severe bleeding occurs.
No routine anticoagulant monitoring is required as INR tests are unreliable for patients on DOACs.

Missed doses

If a dose is missed, follow the guidance below:
Once a day regimen: a missed dose should be taken as soon as possible after it is noticed, but only on the same day. If this is not possible, the patient should skip the dose and continue with the next dose as prescribed. The patient should not take two doses to make up for a missed dose.
Two times a day regimen: a missed morning dose should be taken immediately when it is noticed, and it may be taken together with the evening dose. A missed evening dose can only be taken during the same evening, the patient should not take two doses the next morning.

If a child vomits within 30 minutes of taking a dose, the dose can be repeated.
If the child vomits more than 30 minutes after the dose, do not repeat the dose.

Surgery in patients taking rivaroxaban

Urgent surgery is needed and there is a *high risk of bleeding* – seek advice from the Haematology team. If there is a *low risk of bleeding*, it may be reasonable to undertake surgery without interrupting rivaroxaban administration (except while fasting).
Elective surgery is planned – Fontan and DCM patients will have a low day-to-day risk of acute thrombosis. Rivaroxaban can therefore be stopped prior to elective surgery and there

is likely to be a low benefit in having bridging anticoagulation with unfractionated or low molecular weight heparin.

- Minor surgery with low risk of bleeding: stop rivaroxaban 24 hours prior to surgery (i.e. no dose the day before or the morning of surgery). Restart it after surgery the same or next day when the bleeding risk has passed.
- Major surgery with higher risk of bleeding: stop rivaroxaban 48 hours prior to surgery (i.e. no dose from 2 days prior to the operation). Restart the rivaroxaban 2 days after surgery if there is no other concern about bleeding.
- PICC line placement: check for local guidance

If feeding has not been re-established post-op, consider bridging with LMWH or IV heparin. This decision should be made by the treating physician.
In patients with renal impairment, the rivaroxaban may need to be stopped sooner than the timimgs mentioned above – seek advice from Haemtology.

Bleeding on Rivaroxaban

There is currently no licensed reversal agent for rivaroxaban in children. If a patient on rivaroxaban has a bleeding complication the following advice applies:

- Minor bleeding – stop treatment and investigate source of bleeding
- Major bleeding – discuss with the on-call Haematologist. Discuss use of prothrombin complex concentrate (PCC)* (PCC - Factor II, VII, IX and X concentrate) at a dose of 25-50 U/kg. PCC is obtained from blood bank (not pharmacy) – dose to be advised by haematologist. Dissolve in water for injection as per manufacturer's guidance, using an aseptic technique and the provided transfer device. Administer over 10 minutes.
- Discuss with Haematology about possible use of Andexanet alpha. There are very little data in children, but this may be of some benefit if bleeding continues to be an issue. Also discuss possible use of Octaplex 500 (see octaplex 500 IU powder and solvent for solution for injection - Summary of Product Characteristics (SmPC) - (emc) (medicines.org.uk))
- Please note - Vitamin K will not have any effect on DOAC reversal or levels and would therefore not be recommended.

See:
https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6913453/#:~:text=Bleeding%20is%20the%20main%20complication%20of%20anticoagulant%20therapy&text=2%2D6-,Although%20direct%20oral%20anticoagulants%20(DOACs)%20reduce%20the%20risk%20of%20major,patients%20who%20have%20major%20bleeds.

Rivaroxaban Appendix 1 - Criteria Table

Print this page for the notes – also refer to patent counselling sheet (Rivaroxaban Appendix 2)

Absolute and relative contraindications for cardiac patients	**Y**	**N**
Hepatic disease with coagulopathy or cirrhosis		
Hypersensitivity to drug or excipients		
Other concomitant anti-coagulants (which need to		
Co-existing bleeding risk		
Congenital/acquired bleeding disorder		
Uncontrolled hypertension		
Retinopathy		
Bronchiectasis/pulmonary bleeding		
Other bleeding risk		
Renal impairment (GFR <50)		
Concomitant meds **e.g.** inhibitors of CYP34A or P-gp (example azole antifungals) or inducers of CYP3A4 (rifampicin, phenytoin)		
Metallic heart valve (warfarin preferable)		
Triple positive antiphospholipid syndrome		
Severe hypoalbuminaemia states		
Feeding via naso-jejunal tube or surgical jejunostomy tube		
Cardiac Eligibility Criteria		
Patient with Fontan circulation or dilated cardiomyopathy >12 kg in weight		
Has had 5 days or more of low molecular weight heparin or unfractionated heparin or has been established on warfarin and the INR is <2.5		
Family aware of risks and information sheet given		
Baseline investigations complete and within acceptable range (FBC, U&Es, LFTs, clotting screen)		
Able to take with/after food or milk feed		
Length of treatment agreed, or follow up review arranged		

References:

Rivaroxaban SPmC: https://www.medicines.org.uk/emc/product/12108/smpc

Male C et al. EINSTEIN-Jr Phase 3 Investigators. Rivaroxaban compared with standard anticoagulants for the treatment of acute venous thromboembolism in children: a randomised, controlled, phase 3 trial. Lancet Haematol. 2020 Jan;7(1):e18-e27. doi: 10.1016/S2352-3026(19)30219-4. Epub 2019 Nov 5. PMID: 31699660.

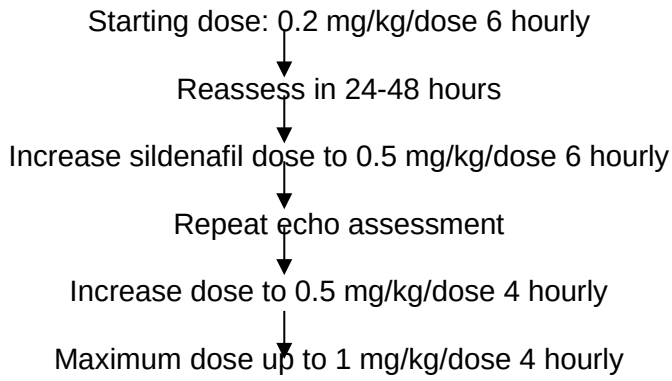
Monagle P, et al. Bodyweight-adjusted rivaroxaban for children with venous thromboembolism (EINSTEIN-Jr): results from three multicentre, single-arm, phase 2 studies. Lancet Haematol. 2019 Oct;6(10):e500-e509. doi: 10.1016/S2352-3026(19)30161-9. Epub 2019 Aug 13. PMID: 31420317.

Connor P et al. Safety and efficacy of rivaroxaban in pediatric cerebral venous thrombosis (EINSTEIN-Jr CVT). Blood Adv. 2020 Dec 22;4(24):6250- 6258. doi: 10.1182/bloodadvances.2020003244. PMID: 33351120; PMCID: PMC7756994

Reviewed 2023 – Authors: Clara Danielsen, Susie Gage, Srinivas Narayan, Jack Gibb, Dirk Wilson and with thanks to Dr Phil Connor

7.17 Sildenafil Guideline

Sildenafil is being used increasingly, particularly in the newborn period (e.g. PPHN and chronic lung disease patients). It should be instituted under consultant direction as follows:



Consider referral to the GOS National Pulmonary Hypertension Service in patients for whom long-term sildenafil therapy is anticipated.

NB – **chronic use** of high dose sildenafil in patients with PA hypertension is associated with worse outcomes and the MHRA has issued the following advice:

For patients who weigh ≤ 20 kg the maximum recommended dose is 10 mg three times a day and for patients > 20 kg the maximum recommended dose is 20 mg three times a day.

Contributors: Orhan Uzun, Clara Danielsen, Dirk Wilson, Amos Wong. Reviewed 2023

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