

Referral Pathway for electrophysiology and device procedures in adult patients with congenital heart

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Target Audience

Physicians or Nurses within the North West and North Wales and the Isle of Man Operational Delivery Network.

Patients affected

Patients ≥ 18 years of age requiring assessment for or undergoing electrophysiology and device procedures

In patients with congenital heart disease (CHD) there are structural and hemodynamic factors to consider when managing arrhythmias and society guidelines recommend referral and discussion of EP and device cases with ACHD specialists prior to intervention. In addition, there may be anatomical and haemodynamic challenges not seen in acquired heart disease requiring that procedures are undertaken by those with expertise in arrhythmias in ACHD.

The majority of patients with congenital heart disease will require lifelong follow up. New onset arrhythmia may signal the need for earlier follow up by the ACHD team or maybe an opportunity for re-referral of patients lost to follow up.

In principle all patients with arrhythmias and congenital heart disease should be referred to the team at LHCH for review and further management. It maybe that some simpler procedures and simple CHD lesions can be managed locally but this should only be after discussion.

Example electrophysiology procedures that maybe performed locally after review by the ACHD team

1. Atrial arrhythmia ablation in patients with isolated congenital aortic or mitral valve abnormalities such bicuspid aortic valve disease or mitral valve prolapse
2. Atrial arrhythmia ablation in patients with an isolated restrictive perimembranous VSD or isolated repaired coarctation
3. Atrial arrhythmia ablation in patients with isolated repaired PDA
4. Atrial arrhythmia ablation with isolated PFO

5. Atrial arrhythmia ablation in patients with isolated mild pulmonary stenosis having never required treatment

Example cardiac device procedures that maybe performed locally after review by the ACHD team

1. Pacemaker generator change
2. Pacemaker insertion in patients with repaired isolated ASD or VSD with no residual shunt
3. Pacemaker insertion in patients with patients with isolated congenital aortic or mitral valve abnormalities such bicuspid aortic valve disease or mitral valve prolapse-repaired or unrepaired
4. Pacemaker insertion in patients with isolated mild pulmonary stenosis having never required treatment
5. Pacemaker insertion in patients with isolated repaired PDA or isolated repaired coarctation

Referral Guidelines

Patients who may be discharged after an initial ACHD review:

- > **Atrial septal defect (ASD)**
 - > with early surgical repair
 - > no residual defect
 - > normal cardiac volumetrics
 - > normal function
 - > normal ECG
- > **Ventricular septal defect (VSD)**
 - > with early surgical closure or spontaneous closure
 - > normal conduction on the ECG
- > **Patients with a Patent Foramen Ovale (PFO)**
- > **Patients with a Patent Ductus Arteriosus (PDA)**
 - > device closure or coiling

Patients who could be followed locally by acquired cardiology with return referral if needed or joint care:

- > small restrictive ventricular septal defect (peri-membranous or muscular)
- > small atrial septal defect (shunt <1.5:1)
- > mild pulmonary stenosis or pulmonary regurgitation
- > bicuspid aortic valve with mild stenosis and/or regurgitation and no aortopathy
- > mild mitral regurgitation

Patients who can be managed jointly in a peripheral clinic with periodic review by an ACHD specialist:

- > late atrial septal defect surgical repair (adult) or device closure (device size >24mm)
- > atrioventricular septal defect with excellent repair
- > all valve replacements or post valvotomy
- > Tetralogy of Fallot with good repair <moderate pulmonary incompetence
- > repaired coarctation of the aorta with excellent repair (simple coarctation)
- > Isolated coarctation of the aorta not requiring intervention and normotensive

Patients who are best cared for by an ACHD cardiologist- centrally or locally:

- > restrictive ventricular septal defect with aortic regurgitation
- > atrioventricular septal defect with >moderate left or right atrioventricular valve regurgitation (LAVVR/RAVVR)
- > Tetralogy of Fallot with >moderate Pulmonary Regurgitation (PR)
- > Sub or supra aortic stenosis
- > Ross repairs
- > Dysplastic aortic valve/Bicuspid Aortic Valve (BAV) with >moderate Aortic Incompetence (AI) or Aortic Stenosis (AS) +/-aortopathy
- > Complex coarctation
- > Truncus arteriosus
- > Shone's complex
- > All patients with a history of transposition of the great arteries (Mustard, Senning, Rastelli, Arterial Switch)
- > Pulmonary atresia with or without ventricular septal defect
- > Ebstein's anomaly
- > All Fontan patients
- > Any unrepaired single ventricle physiology
- > Patients with ACHD-PH and Eisenmenger's syndrome

List of ACHD Clinics across the North West of England and North Wales Table 1:

Hospital	Contact	Telephone Number
Liverpool Heart & Chest Hospital NHS Foundation Trust Thomas Drive Liverpool L14 3PE	ACHD Cardiologists: Dr Damien Cullington (Clinical Lead) Dr Sarah Elgamal (imaging) Dr Jaspal Dua Dr Petra Jenkins (PH) Dr Reza Ashrafi (EP) Dr Vasileios Papaioannou (PH)	Switchboard: 0151 600 1616

	Dr Julia Jones	
	Specialist Nurses - Adult Congenital Heart Disease	Tel: 0151 254 3333 Mobile: 07542229889
Manchester University NHS Foundation Trust, Manchester Heart Centre, MRI, Oxford Road, Manchester, M13 9WL	ACHD Cardiologists Dr Damien Cullington (Clinical Lead), Dr Jaspal Dua Dr Petra Jenkins (PH) Dr Vasileios Papaioannou (PH and general ACHD) Dr Julia Jones	Switchboard: 0161 276 1234
	Cardiologists with a special interest in ACHD Professor Bernard Clarke	
	Specialist Nurses - Adult Congenital Heart Disease	0161 276 1234 Bleep: 6994 Voicera: ask for ACHD Nurse
Manchester University NHS Foundation Trust, Wythenshawe Hospital, Southmoor Road, Wythenshawe M23 9LT	Consultant Cardiologist with an interest in ACHD Dr Anita Macnab ACHD Cardiologist Dr Reza Ashrafi	Tel: 0161 998 7070
	Specialist Nurses - Adult Congenital Heart Disease	0161 276 1234 Bleep: 6994 Voicera: ask for ACHD Nurse
Betws Cadwalader University Health Board – Wrexham Maelor Hospital, Croesnewydd Road, Wrexham, LL13 7TD	Consultant Cardiologist with an interest in ACHD Dr Raj Thaman ACHD Cardiologist Dr Petra Jenkins (Based at LHCH)	Tel: 01978 727 801
	Nicola Coates Specialist Nurse - Adult Congenital Heart Disease	Tel: 01978 727 801
Blackpool Teaching Hospitals NHS Foundation Trust,	ACHD Cardiologist Dr Vasileios Papaioannou (Refer to LHCH FAO of Blackpool ACHD)	Tel: 0151 254 3350

Blackpool Victoria Hospital, Whinney Heys Road, Blackpool, FY3 8NR	Specialist Nurses – Adult Congenital Heart Disease via the helpline at Liverpool Heart & Chest Hospital	Tel: 0151 254 3333
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How to refer

All new diagnoses of congenital heart disease should be referred in writing to a member of the level 1 team either centrally at Liverpool Heart and Chest NHS Foundation Trust or at the peripheral clinic that they visit. Patients will either then receive an appointment or advice will be given back to the referrer suggesting local follow up.

Patients who have been identified as being lost to follow up should be referred in writing to a member of the level 1 team either centrally at Liverpool Heart and Chest NHS Foundation Trust or at the peripheral clinic that they visit. Patients will either then receive an appointment or advice will be given back to the referrer suggesting local follow up.

Acute admissions can be arranged via the consultant on-call via switchboard at Liverpool Heart and Chest NHS Foundation Trust on 0151 600 1616.

Who to Contact

The ACHD on-call team are available for urgent advice twenty-four hours a day, seven days a week and can be contacted via the switchboard at Liverpool Heart and Chest NHS Trust on 0151 600 1616.

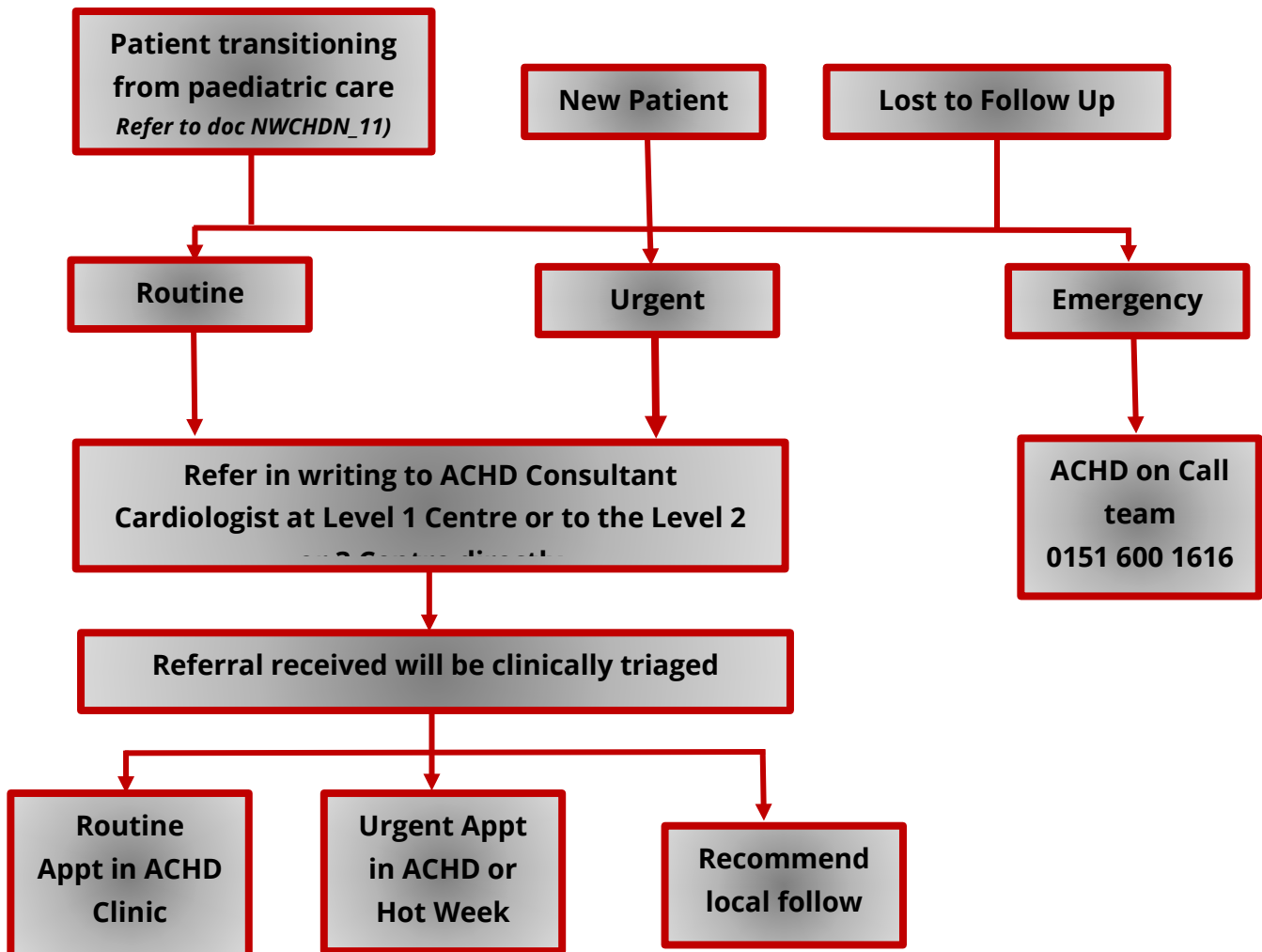
Dr Reza Ashrafi
 Dr Damien Cullington
 Dr Jaspal Dua
 Dr Sarah Elgamal
 Dr Petra Jenkins
 Dr Vasileios Papaioannou
 Dr Julia Jones

**ACHD Specialist nursing team can be contacted Monday to Friday 8am to 6pm
07542229889**

Fiona Andrews
Lisa Devitt
Lorraine English
Anna Harrison
Sharon Hughes
Julie Ollerhead
Linsha George

Diagram 1

ACHD Referral Pathway



References

2018 AHA/ACC guidelines for the management of adults with congenital heart disease
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2014 AHA/ACC Guideline for the Management of Patients With Valvular Heart Disease
A Report of the American College of Cardiology/American Heart Association Task Force
on Practice Guidelines
<http://www.onlinejacc.org/content/63/22/e57>

2014 PACES/HRS Expert Consensus Statement on the Recognition and Management of
Arrhythmias in Adult Congenital Heart Disease
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2020 ESC Guidelines for the management of adult congenital heart disease: The Task
Force for the management of adult congenital heart disease of the European Society of
Cardiology (ESC)
[https://academic.oup.com/eurheartj/advance-
article/doi/10.1093/eurheartj/ehaa554/5898606](https://academic.oup.com/eurheartj/advance-article/doi/10.1093/eurheartj/ehaa554/5898606)

2017 ESC/EACTS Guidelines for the management of valvular heart disease
<https://academic.oup.com/eurheartj/article/38/36/2739/4095039#supplementary-data>

2018 ESC Scientific Document Group, Arrhythmias in congenital heart disease: a
position paper of the European Heart Rhythm Association (EHRA), Association for
European Paediatric and Congenital Cardiology (AEPC), and the European Society of
Cardiology (ESC) Working Group on Grown-up Congenital heart disease, endorsed by
HRS, PACES, APHRS, and SOLAECE
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patients-with-congenital-heart-disease.pdf](https://bhrc.com/wp-content/uploads/2019/03/160216-Standards-Cardiac-rhythm-management-in-patients-with-congenital-heart-disease.pdf)