BRITISH CONGENITAL CARDIAC ASSOCIATION

REFERRAL GUIDELINES FOR ACCESSING CONGENITAL CARDIAC SERVICES FETAL, PAEDIATRIC AND ADULT

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These guidelines should be read in conjunction with the NHS England congenital heart disease standards and specifications published in 2016[1]. They are derived from multiple sources[2,3] and use the following definitions[1]:

ACHD services
Level 1 Specialist ACHD surgical centre
Level 2 Specialist ACHD centre
Level 3 Local ACHD centre

Paediatric cardiac services
Level 1 Specialist Children's Surgical Centres
Level 2 Specialist Children's Cardiology Centres
Level 3 Local Children's Cardiology Centres

Fetal cardiac services
Generally based at Level 1 and 2 congenital units

Referral pathways

Emergency
Emergency referral after assessment and initial management in the local Level 3 hospital should be directly to the:
1 Regional paediatric/ ACHD cardiac centre (Levels 1 and 2) and
2 Regional transport service and
3 Regional paediatric/ adult cardiac ICU

Acute
There will be a group of patients who present to local Level 3 hospitals who are not sick enough to require emergency transfer but in whom the local team (consultant or registrar) wish for advice/ transfer to the Level 1 or 2 centre/ urgent outpatient assessment. These patients should be discussed with either the on call consultant or on call registrar/ SpR/ fellow at the Level 1/ 2 centre depending on local arrangements. Acute fetal referrals should be discussed by the obstetricians or antenatal sonographers directly with the fetal cardiology team at the Level 1 or 2 centre.

Routine
These should be considered for assessment by a paediatrician, paediatrician with expertise in cardiology (PEC), adult cardiologist or adult cardiologist with expertise in congenital cardiology in a Level 3 clinic, before being referred to the Level 1 or 2 centre if needed. Routine fetal referrals from the obstetricians or antenatal sonographers should be to the fetal cardiology team at the Level 1 or 2 centre.

Types of patients seen
Patients generally belong to the following categories:
- Prenatal diagnosis
- Follow-up of previously diagnosed congenital heart defects
- Follow-up management of arrhythmias
- Follow-up of post-operative cardiac surgical or intervention patients
- Referrals from level 1/ 2 centres outside the network to level 3 centres in the network often for more local cardiac investigations eg Holters etc
- Referrals from GPs, paediatricians, community paediatricians, adult cardiologists and other specialities.

Antenatal diagnosis
Referrals are generally from obstetric teams or antenatal ultrasonographers. Patients are referred for the following indications:
- Maternal indications e.g. maternal congenital heart disease, metabolic disorder, exposure to teratogen, maternal collagen disease with anti-Ro/ SSA and/ or La/ SSB, NSAIDs exposure after 25 weeks gestation
- Familial indications e.g. family history congenital heart disease, genetic disorders, chromosomal abnormalities, syndromes associated with congenital heart disease or cardiomyopathy
- Fetal indications e.g. suspicion of fetal cardiac abnormality, nuchal translucency >3.5mm at first trimester screening, nuchal pad thickening >6mm at mid-trimester anomaly screening, fetal hydrops, pericardial effusion, pleural effusion, extra-cardiac malformation associated with congenital heart disease, chromosomal abnormality/ genetic syndrome associated with congenital heart disease, fetal arrhythmia, fetal conditions associated with fetal cardiac failure e.g. twin-twin transfusion syndrome, absence of ductus venosus, fetal anaemia, cardiac tumour, arteriovenous fistulae, acardiac twin.

If an antenatal diagnosis of a congenital heart defect has already been made, a plan for the most appropriate site for delivery and immediate plan for treatment (e.g. prostaglandin) should have been made by the fetal cardiologist.

Neonates and young infants up to 8 weeks

Newborn with a murmur and otherwise clinically well:

1. These babies need a review before discharge by a senior neonatologist or paediatrician, including saturation monitoring. If the murmur is assessed as representing a significant risk of congenital heart disease, or there are other concerns such as desaturation (<95%), then the case will need to be discussed urgently with the paediatric cardiology team at the network Level 1 or 2 centre.

2. Babies with a murmur should be reviewed within 2 weeks in the neonatal clinic including saturation monitoring unless an experienced clinician can confidently assess them as being at low risk of having significant underlying congenital heart disease, in which case outpatient follow-up should be arranged in 4 to 6 weeks.

A verbal explanation and in addition a patient leaflet should be given to parents of all babies with a murmur. Clear instructions and guidance should be given to the parents including red flags that clarify when to seek help and prompt medical review and when to seek help with their GP or at A&E.

3. If there is a persistent murmur that is considered consistent with congenital heart disease at the 2-week review, then the baby should be booked into the local Level 3 paediatric cardiology clinic. The baby should follow the pathway outlined in the Appendix.

If the echocardiogram performed at the Level 3 centre shows a cardiac defect unlikely to be haemodynamically significant, such as a small atrial septal defect, patent arterial duct or small ventricular septal defect, or a minor degree of outflow obstruction, then the baby should be reviewed in 4 to 10 weeks in the Level 3 clinic.

Neonates and infants diagnosed with significant congenital heart defects will require individual discussion between Levels 3 and 1/ 2 on a case-by-case basis.

New referrals of older infants (>8 weeks) and children (generally <16 years but sometimes <18 years)

These referrals generally fall into the following categories:
1. Murmur significant enough to require diagnostic evaluation
2. Cyanosis
3. Chest pain
4. Palpitations
5. Syncope or dizziness
6. Referral for screening because of family history of congenital heart defect, inherited cardiac condition such as cardiomyopathy or other syndromes
7. Kawasaki disease

These patients should be seen initially by a paediatrician with expertise in paediatric cardiology (PEC):

1. ***Murmur:*** This guidance should not exclude general paediatricians from assessing the significance of the murmur. If the murmur is suspected of being pathological or an abnormality found on an echocardiogram performed locally, then the child should be referred to the local PEC at the Level 3 centre as shown in the Appendix. Minor lesions, such as small ventricular septal defects, atrial shunts, or mild valvar stenosis may be followed up there as appropriate. Persisting lesions should be reviewed within 1 year by the visiting paediatric cardiologist.

2. Cyanosis: Haemoglobin oxygen saturation should be checked. If there is evidence of central cyanosis, then the patient should be discussed with the local PEC, or if they are not available with the on call paediatric cardiology team at the Level 1 or 2 centre.

3. Chest pain: Generally these patients and their parents require reassurance as to the musculo-skeletal nature of the chest pain. The patients can be seen in a general paediatric clinic. If a cardiac opinion is required, then they should be seen by the local PEC in clinic and investigations arranged if deemed necessary. If these are normal the patient/ parents can be reassured and discharged.

4. Palpitations: These patients should be seen in the general paediatric clinic and investigations arranged. These may include ECG, 24- or 48-hour ECG tape, event recorder and Alivecor device if available and if indicated an echocardiogram and treadmill exercise test. If all these investigations are normal, the patient could be discharged. If there is still doubt or if there is an abnormality found, then the patient should be discussed with the local PEC, or if they are not available, the paediatric cardiologist.

5. Syncope or dizziness: Patients should be seen by a paediatrician in the general paediatric clinic initially to ascertain whether the symptoms are vasovagal, arrhythmic, neurological or other in origin and appropriate investigations arranged.

6. Referral for family history of congenital heart defects, cardiomyopathy, and other syndromes: Services for Inherited Cardiac Conditions (ICC) are usually provided by a specialised service at the Level 1 or 2 centre. These families can be discussed with the ICC service at the discretion of the paediatrician, rather than being referred to the paediatric cardiologist.

7. Kawasaki disease:
The guidelines published in the AHA scientific statement in 2017 should be consulted[4]

New referrals of adults (>16-18 years) with suspected or actual congenital disease

These referrals also generally fall into the following categories:
1. Murmur significant enough to require diagnostic evaluation
2. Cyanosis
3. Chest pain
4. Palpitations
5. Syncope or dizziness
6. Referral for screening because of family history of congenital heart defect, inherited cardiac condition such as cardiomyopathy or other syndromes
7. Previous Kawasaki disease
8. Pregnancy with maternal heart disease

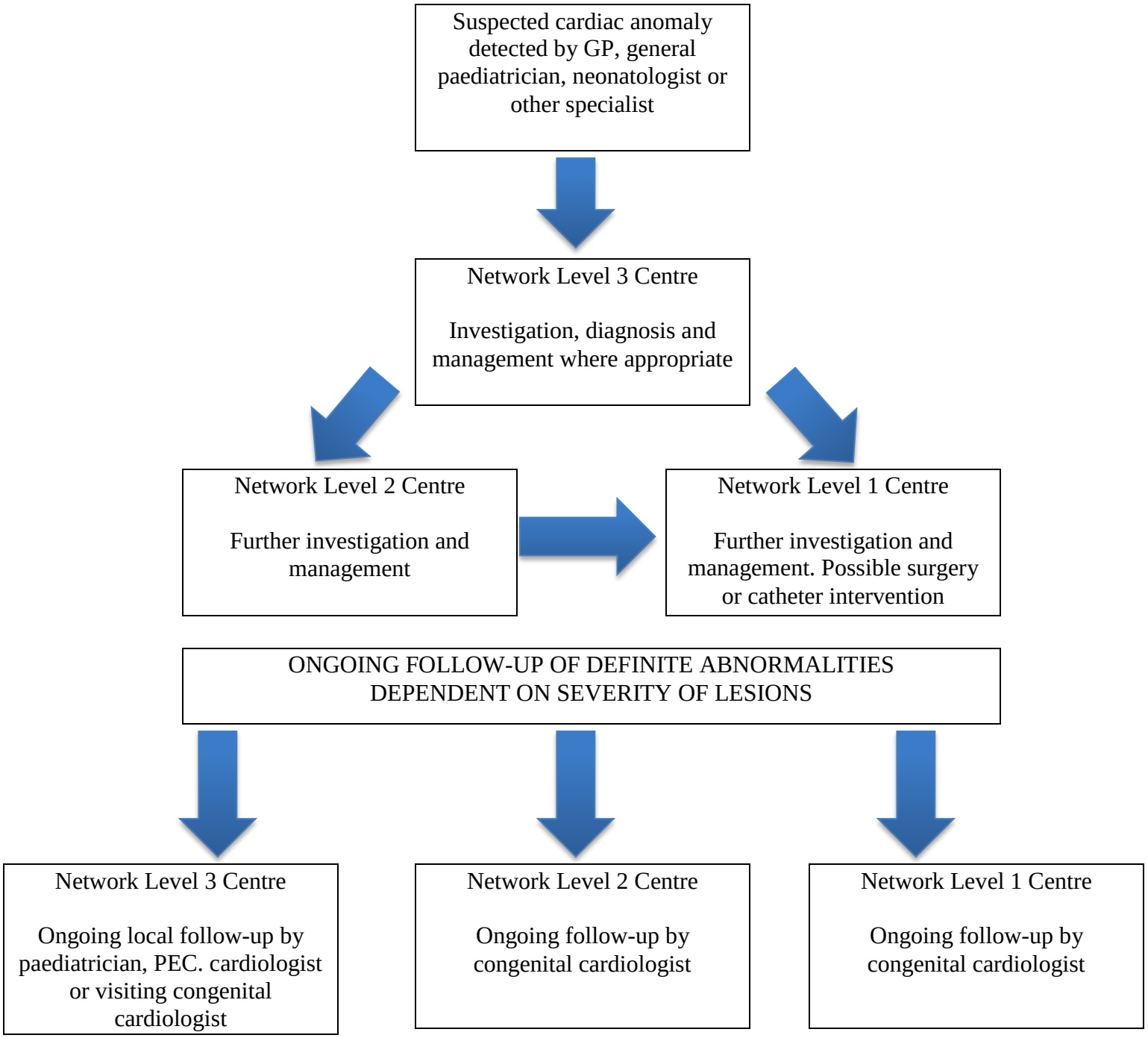
These patients are usually seen locally by the adult cardiologist in the first instance. Where there is definite but minor congenital cardiac disease such as small ventricular septal defects, atrial septal defects, patent foramen ovale or mild valvar stenosis then local follow-up is appropriate. Where there is a more major congenital lesion then liaison with the Level 1 or 2 ACHD cardiology team is necessary.

Children, adolescents and adults diagnosed with congenital heart defects: ongoing follow-up

Suitable follow-up for these patients will be decided jointly between the paediatric/ ACHD cardiologist and the local paediatrician/ cardiologist. Follow-up will be dependant on the individual patient and may take place in the Level 1, 2 or 3 centre and often as shared care.

Appendix

Referral pathway for children and adults with suspected cardiac abnormality



Acknowledgements
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References

1 Congenital heart disease standards & specifications. NHS England 2016. https://www.england.nhs.uk/wp-content/uploads/2018/08/Congenital-heart-disease-standards-and-specifications.pdf

2 BCCA outreach clinic paediatric cardiology service 2009. http://www.bcs.com/documents/9ZD_Outreach_clinic_Paediatric_Cardiology_Service_BCCA_document_Oct_2009.pdf

3 Referral guidelines for paediatric cardiology outpatient clinics. Yorkshire and Humber congenital cardiac network 2017. https://www.networks.nhs.uk/nhs-networks/yorkshire-and-humber-congenital-cardiac-network/network-board-meetings/february-2019-agenda-and-papers/paediatric-referral-guidelines/file_popview

4 Diagnosis, treatment, and long-term management of Kawasaki disease: A scientific statement for health professionals from the American Heart Association. McCrindle et al 2017. https://doi.org/10.1161/CIR.0000000000000484