# Congenital heart disease

## *Executive summary*

## Introduction

Congenital heart defects (CHD) are problems with the structure of the heart that are present at birth. It is caused by abnormal formation of the heart during foetal development. In most cases when a baby is born with a congenital heart defect, there is no known cause for it. CHD are classified into two broad categories, acyanotic (ACHD) and cyanotic lesions (CCHD). The most common acyanotic lesions are ventricular septal defect (VSD), atrial septal defect (ASD), atrioventricular canal defects and patent ductus arteriosus (PDA). In patients with Cyanotic congenital heart defects, the blood that is pumped around the body contains less than normal levels of oxygen causing the child to have bluish tinge to the skin and mucous membranes. Tetralogy of Fallot is the most common cyanotic heart defect but may not always become apparent immediately after birth.

CHDs are the most common cause of infant death due to birth defects. Approximately 25% of children born with a CHD will need heart surgery or other interventions to survive. MRCG has an ongoing collaboration with the Chain of Hope charity which assist in management of children with CHD, by making arrangements for surgical treatment overseas for those children who will benefit. Please review the current SOP or discuss with a consultant if you need more advice about how to ensure children with CHD are included in this programme.

## Target users

* Doctors
* Nurses

## Target area of use

* Outpatient department
* Ward

## Key areas of focus / New additions / Changes

This guideline outlines the diagnosis and management of congenital heart defects.

## Limitations

Patients with congenital heart disease need to be referred overseas for surgical repair. This is best achieved through ongoing care at our clinic and attendance at our periodic screening clinic (about once per year) run by Chain of Hope, who can arrange sponsorship for eligible children.

## Presenting symptoms and signs

* Failure to thrive (Small for age)
* Feeding difficulties
* Diaphoresis
* Respiratory distress
* Cyanosis (in cyanotic CHD)
* Cardiac murmurs
* Detection of a heart murmur - need to differentiate innocent from pathological
* Shock - when duct closes in severe left heart obstruction

## Examination findings

Full cardiac and respiratory examination is important, including basic observations, inspection, palpation and auscultation.

Pulse oximetry is helpful to identify oxygen saturations.

## Investigations

ECHO is the gold standard for making the diagnosis.

Other tests that might be helpful include:

* CXR – to see if the heart is enlarged or if there are signs of left heart failure.
* ECG – to identify rhythm problems.
* Cardiac catheterization is not available in Gambia – but can help determine the function of the heart valves and chambers.

## Management

The management of CHD depends on the exact problem identified – these are described below.

### Asymptomatic Child With a Systolic Murmur

When murmur is identified in an asymptomatic child 2 years of age or older, the child should be evaluated with full history, examination, CXR (and perhaps ECG).

ECHO is reserved for selected cases where the history and physical findings are atypical for an innocent murmur. Once a murmur is determined to be innocent, further cardiac evaluation is unnecessary.

### Ventricular Septal Defect

#### Small VSD

In most patients, a small VSD is a relatively benign condition. It is important to monitor these patients to detect and manage the relatively rare but important complications of endocarditis, arrhythmias, aortic regurgitation and subvalvular pulmonary stenosis and discrete subaortic stenosis.

Clinical examination and ECHO are adequate to establish the diagnosis.

Children with a small VSD are often asymptomatic and have an excellent long-term prognosis. Neither medical therapy nor surgical therapy is indicated. Prophylactic antibiotic therapy against endocarditis is no longer indicated in most cases.

#### Moderate to large VSD

In moderate or large VSDs, medical therapy is indicated to manage symptomatic congestive heart failure (CHF). This may include:

* Increase feeds to ensure adequate weight gain. Occasionally, oral feeds must be supplemented with nasogastric tube feedings, because a baby in CHF may be unable to consume adequate calories for appropriate weight gain.
* Furosemide to relieve pulmonary congestion – give 1-3 mg/kg/day divided into 2 or 3 doses. Long-term furosemide treatment may result in hypercalciuria, renal damage, and electrolyte disturbances.
* ACE inhibitors (eg captopril and ramipril) reduce both the systemic and pulmonary pressures (the former to a greater degree), thereby reducing the left-to-right shunt.

Uncontrolled CHF with growth failure and recurrent respiratory infection is an indication for immediate surgical repair. Neither the age nor the size of the patient is prohibitive in considering surgery.Ideally, surgery is performed at 3 – 6 months of age to prevent permanent lung damage from pulmonary hypertension.

### Patent Ductus Arteriosus (PDA)

In most cases, Doppler ECHO is adequate to confirm the diagnosis. Cardiac catheterization is necessary only in atypical cases.

Treatment is by elective closure of the PDA.

When the duct is large complications include infective arteritis, aneurysm of the ductus, congestive heart failure, pulmonary vascular obstructive disease, and calcification of the ductus.

### Atrial Septal Defect (ASD)

There are two main types of ASD – secundum ASD (80% of ASD) and partial atrioventricular septal defect AVSD or primum ASD. Both present with similar symptoms and signs but their anatomy is different.

Patient may have no symptoms or may present with recurrent chest infections and wheeze. ECHO is the mainstay of diagnosis.

Once an ASD is found at ECHO, an assessment of shunt size is made.

* Small shunt: Absence of diastolic flow rumble, normal chest X-ray, ECHO evidence of normal right ventricular size and normal interventricular septal motion or a pulmonary to systemic flow ratio (QP/QS) <1.5.
* Large shunt: Presence of a diastolic flow rumble or ECG evidence of right ventricular hypertrophy, CXR evidence of cardiomegaly or increased pulmonary vascular markings, or ECHO evidence of right ventricular enlargement or paradoxical septal motion or a QP/QS 1.5.

Many small ASDs close spontaneously below the age of 2 years and some up to the age of 4 years, therefore conservative treatment is indicated if the patient is under age 4 years and asymptomatic. Even those with symptoms and a large left-to-right shunt can be observed up to the age of 5 years.

If the ASD does not close spontaneously, then surgical closure is indicated at 3-5 years of age in order to prevent right heart failure and arrhythmias in later life.

### Tetralogy of Fallot (TOF)

TOF is the most frequent cyanotic congenital heart disease. The four cardinal anatomic features are a large VSD, overriding of the aorta, subpulmonary stenosis causing right ventricular outflow tract obstruction (RVOTO) and right ventricular hypertrophy. The severity of RVOTO determines the extent of R-L shunting on ventricular level and therefore the extent of cyanosis.

* **Severe RVOTO** results in deep cyanosis due to a large R-L shunt.
* **Mild RVOTO** does not cause cyanosis (“pink Fallot”) and may result in heart failure due to large L-R shunting on ventricular level

*Examination findings*

Central cyanosis and a systolic murmur, caused by PS with the intensity inversely correlated with severity of PS. The VSD is not associated with a murmur!

*Investigations*

Echocardiography is the cornerstone for the diagnosis. Chest X-ray and ECG may be helpful.

Pulse oximetry at each clinical visit. If O2 saturations repeatedly < 80%, treatment is mandatory.

*Management of cyanotic spells*

These are caused by acute obstruction of RVOT or acute decrease of peripheral resistance.

They present with:

* Increase of cyanosis
* Tachycardia
* Auscultation decreased intensity of systolic heart murmur due to reduced flow into PA
* Seizures (neurological sequelae)

Older untreated children have signs of chronic hypoxaemia including digital clubbing. They are at risk of endocarditis and brain abscess and these risks increase with age, raised Hb and iron deficiency.

*Emergency treatment*

* Increase peripheral resistance by encouraging knee-to-chin position
* Administer oxygen
* Sedation: Morphine sc or iv 0.1-0.2 mg/kg OR Midazolam iv 0.1 mg/kg OR Midazolam Rectal 0.5 mg/kg OR Ketamine iv 1-2 mg/kg OR Ketamine im 5 mg/kg.
* Give fluid: iv bolus of 10 ml/kg of normal saline which can be repeated.
* Give betablocker: iv propranolol 15-20 mcg/kg – repeat up to a maximum of 100 mcg/kg. Once child is improving, continue with doses of 15-20 mcg/kg up to 6 hourly until able to take it orally.

*Ongoing treatment*

* Oral propranolol 2-6 mg/kg/day in divided doses.
* Iron replacement as despite the plethora, children with TOF are often iron deficient.

Surgical repair involves enlargement of RVOT and VSD closure.

### Children who have had surgery for CHD

Review patients in clinic about 3 months after surgery. Check for resolution of all post-operative problems. Examine the patient fully.

Look for important complications such as arrhythmia and pulmonary valve insufficiency.

ECHO to confirm successful treatment is usually done by the surgeons, but may need to be repeated here.

ECG, CXR and other tests to assess the patient may be needed if there are any abnormal clinical findings.

### Antibiotic prophylaxis for dental procedures

Many patients with congenital cardiac defects require prophylaxis for bacterial endocarditis when undergoing certain surgical and dental procedures. Antibiotics before dental procedures are only recommended for patients with the highest risk of infective endocarditis:

* A prosthetic heart valve or who have had a heart valve repaired with prosthetic material.
* A history of endocarditis.
* A heart transplant with abnormal heart valve function
* Certain congenital heart defects including:
  + CCHD that has not been fully repaired, including children who have had a surgical shunts and conduits.
  + CHD that's been completely repaired with prosthetic material or a device for the first six months after the repair procedure.
  + Repaired CHD with residual defects, such as persisting leaks or abnormal flow at or adjacent to a prosthetic patch or prosthetic device.

## References

## Lissauer, T., Carroll, W., & Craft, A. (2018). *Illustrated textbook of paediatrics*. Fifth Edition. Elsevier

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