

CONGENITAL HEART DISEASE

SPECIFIC MANAGEMENT STRATEGIES FOR COMMON LESIONS

LEFT TO RIGHT SHUNTS

Atrial septal defects (ASD)

- *small defects*: - no treatment
- *large defects*: - elective closure at 4-5 years age

Ventricular septal defects (VSD)

- *small defects*:
 - no treatment; high rate of spontaneous closure
 - SBE prophylaxis
 - yearly follow up for aortic valve prolapse, regurgitation
 - surgical closure indicated if prolapsed aortic valve
- *moderate defects*:
 - anti-failure therapy if heart failure
 - surgical closure if:
 - heart failure not controlled by medical therapy
 - persistent cardiomegaly on chest X-ray
 - elevated pulmonary arterial pressure
 - aortic valve prolapse or regurgitation
 - one episode of infective endocarditis
- *large defects*:
 - early primary surgical closure
 - pulmonary artery banding followed by VSD closure in multiple VSDs

Persistent ductus arteriosus (PDA)

- *small PDA*:
 - no treatment if there is no murmur
 - if murmur present: elective closure as risk of endarteritis.
- *moderate to large PDA*:
 - anti-failure therapy if heart failure
 - timing, method of closure (surgical vs transcatheter) depends on symptom severity, size of PDA and body weight

Atrioventricular septal defects (AVSD)

- *partial AVSD*:
 - elective surgical repair at 4 to 5 years old;
 - earlier if symptomatic or severe AV valve regurgitation
- *complete AVSD*:
 - primary surgical repair < 6 months age
 - to prevent pulmonary vascular disease
 - In selected patients - e.g. with severe AV valve regurgitation and older patients, conservative treatment is an option as surgical outcomes are poor

OBSTRUCTIVE LESIONS

Pulmonary stenosis (PS)

- *mild*:
 - no treatment
 - (*peak systolic gradient < 50 mmHg*)
- *moderate-severe*:
 - transcatheter balloon valvuloplasty is treatment of choice
 - (*gradient > 50 mmHg*)
- *neonatal critical PS*:
 - characterized with cyanosis and RV dysfunction
 - temporary stabilization with IV Prostaglandin E infusion
 - early transcatheter balloon valvuloplasty

SBE prophylaxis is indicated in all cases

Coarctation of the aorta (CoA)

- *neonatal severe CoA*: frequently associated with large malaligned VSD and intractable heart failure
 - sick infants require temporary stabilization mechanical ventilation
 - correction of metabolic acidosis, hypoglycaemia, electrolyte imbalances
 - IV Prostaglandin E infusion
 - early surgical repair (single-stage CoA repair + VSD closure or 2 stage CoA repair followed by VSD closure at later date)
- *asymptomatic / older children with discrete CoA*:
 - presents with incidental hypertension or heart murmur
 - choice of treatment (primary transcatheter balloon angioplasty, stent implantation or surgical repair)
 - depends on morphology of CoA and age of presentation

CYANOTIC HEART LESIONS

Tetralogy of Fallot (TOF)

- most TOFs suitable for single stage surgical repair at 1 to 2 years age
- indications for modified Blalock Taussig shunt:
 - hypercyanotic spells or severe cyanosis < 6 months age when child is too young for total repair
 - small pulmonary arteries; to promote growth before definitive repair
 - anomalous coronary artery crossing in front of right ventricular outflow tract - precludes transannular incision; repair with conduit required at later age
- following surgical repair, patients need life-long follow up for late right ventricular dysfunction; some may require pulmonary valve replacement

Transposition of the great arteries (TGA)

- *simple TGA*:
 - IV Prostaglandin E infusion promotes intercirculatory mixing at PDA
 - early balloon atrial septostomy (BAS) if restrictive interatrial communication
 - surgical repair of choice: arterial switch operation at 2 to 4 weeks age
 - left ventricular regression may occur if repair not performed within 4 weeks of life
- *TGA with VSD*:
 - do not usually require intervention during early neonatal period; may develop heart failure at 1 to 2 months age
 - elective one-stage arterial switch operation + VSD closure before 3 months age
- *TGA with VSD and PS*:
 - Blalock Taussig shunt during infancy followed by Rastelli repair at 4 to 6 years age

Pulmonary atresia with intact ventricular septum

- IV prostaglandin E infusion to maintain ductal patency in early neonatal period
- further management strategy depends on the degree of right ventricular hypoplasia

Truncus arteriosus

- surgical repair (VSD closure and RV-to-PA conduit) before 3 months of age

Tetralogy of Fallot with pulmonary atresia

- IV prostaglandin E infusion is often required during early neonatal period
- further management strategy depends on the anatomy of the pulmonary arteries and presence of aortopulmonary collaterals

Single ventricle

includes 3 main categories of lesions:

- *double inlet ventricles*:
 - double inlet left ventricle, double inlet right ventricle
- *atretic or stenosed atrioventricular connections*:
 - tricuspid atresia, mitral atresia, hypoplastic left heart syndrome
- *miscellaneous lesions which preclude biventricular circulation*:
 - unbalanced AV septal defect, double outlet right ventricle with remote VSD, congenital corrected transposition of great arteries, heterotaxy syndromes

Requires staged management approach for eventual Fontan procedure

Total anomalous pulmonary venous drainage

- 4 major anatomic types: supracardiac, cardiac, infracardiac and mixed
- management strategy depends on presence of pulmonary venous obstruction
 - *obstructed pulmonary venous drainage (frequent in infracardiac type)*
 - presents with respiratory distress and heart failure
 - initial stabilization: oxygen, diuretics, positive pressure ventilation
 - surgical repair immediately after initial stabilization
 - *unobstructed pulmonary venous drainage*
 - early surgical repair is required