CONGENITAL CARDIAC DISEASE

This may be classified as cyanotic or acyanotic, depending on the presence of central cyanosis. Those with cyanosis will have a right-to-left shunt, preventing complete oxygenation of systemic arterial blood. Some patients with high-flow left-to-right shunts develop severe pulmonary hypertension as a consequence of massive pulmonary blood flow. This result in pressure in the right heart champers that greater than those in the left heart and, consequently, in reversal of the shunt direction to right to left, causing cyanosis. This situation is called Eisenmenger`s syndrome. Primary repair is usually advised for congenital defects, but in some instances it may be helpful to delay definitive repair until the child is older, larger and fitter.

Atrial septal defect

This is the most common abnormality, causing a left to right atrial shunt and hence an increase in right heart and pulmonary blood flow. Patients may be asymptomatic or may present with frequent chest infections. There is a fixed split second heart sound and a pulmonary ejection systolic murmur. Small defects are of little hemodynamic significance, but if the pulmonary to systemic flow ratio exceeds 2:1, closure is necessary. ECG frequently demonstrates right ventricular hypertrophy and echocardiography is diagnostic.

Surgical repair in children carries a low mortality (<1%), but adults presenting with pulmonary hypertension are at greater risk (10%).

Ventricular septal defect

Many ventricular septal defects are small and close within the first year of life. Larger lesions cause a left to right shunt and pulmonary congestion. Defect may be subdivided according to their embryological origins, but most (85%) occur in the membranous septum. Infants with large defects may present with frequent respiratory infections but patients are often asymptomatic. A pansystolic murmur is audible, maximal at the left sternal edge. The second heart sound may be loud. Biventricular hypertrophy is present on ECG and pulmonary plethora may be noted on chest X-ray. Echocardiography is diagnostic. Asymptomatic defects are observed, but early operation is preferred for larger defects to prevent irreversible pulmonary hypertension. Repair is undertaken using a patch, with an operative mortality of 3-5%.

Patent Ductus Arteriosus

If the ductus arteriosus fail to close after birth, pulmonary blood flow is abnormally high, producing pulmonary congestion and hypertension. Infants have retarded growth and a continuous machinery murmur is audible over the precordium and back. The X-ray shows pulmonary congestion, and echocardiography can exclude concurrent intracardiac defect(s). In premature children, the duct may close with an indometacin infusion (prostaglandin E1 inhibition), but clipping or division at left thoracotomy is likely to be needed. Endovascular closure is an option in older children. The operative mortality is low in older children (<1%) but high (25%) in preterm infants, who are generally very unwell.

Coarctation of Aorta

This condition is caused by a narrowing of the thoracic aorta, usually at the level of the ligamentum arteriosum. The lower body is perfused via extensive chest wall collaterals. Upper body hypertension develops, partly due to relative renal hypoperfusion, and may lead to heart failure in infancy. Untreated adults develop hypertensive cerebrovascular and renal problems and accelerate coronary atheroma. Most children and young adults are asymptomatic and present with high blood pressure or an abnormal chest X-ray. The femoral pulses may be impalpable or weak and delayed, and a systolic murmur may be audible over the back. Left ventricular is seen on the ECG and the chest X-ray shows an enlarged heart, reduced aortic knuckle and characteristic rib notching caused by enlarged and tortuous intercostal arteries eroding the ribs near the posterior angles. Balloon angioplasty has been used to dilate the some coarctation in infants, but surgical correction is required. In infants, an onlay patch graft created from the left subclavian artery is used. This has the advantage of growing with the child, although the left arm growth is slightly decreased. Older children and adults are usually managed with Dacron bypass graft. Surgical correction tends to reduce upper body hypertension in children. It is less effective in adults but pharmacological control of hypertension become more reliable.

Tetralogy of Fallot

This is the most common cause of cyanotic congenital heart disease; it comprises a high ventricular septal defect, an aorta that tends to overlie the interventricular septum, pulmonary valvular and subvalvular stenosis, and right ventricular hypertrophy. Right ventricular outflow obstruction causes cyanosis as a result of right-to-left shunting across the ventricular septal defect. Clinical features depend upon the severity of obstruction. This may not be significant when the child at rest but it may be precipitated by adrenergic events, as these increase the subvalvular obstructive effect of hypertrophied right ventricular muscle. Consequently, the child may become blue and faint during feeding or crying. Right ventricular hypertrophy is found on ECG and the pulmonary artery shadow is small in chest X-ray. Echocardiography is diagnostic but complemented by right ventricular angiography, which demonstrates the pulmonary arterial tree. Correction entails closing the ventricular septal defect with patch, resecting the muscle band contributing to the right ventricular out flow obstruction, and enlarging the right ventricular out flow tract with patch placed across the pulmonary valve annulus and along the pulmonary artery if necessary. In those not fit for this procedure or in those with very small pulmonary vessels, a shunt (usually subclavian artery to pulmonary artery, Blalock-Taussig) is created to increase pulmonary blood flow and, helpfully, lead to further pulmonary arterial growth. Definitive correction may then be possible at a later stage.