Acyanotic congenital heart disease

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- Patent ductus arteriosus The ductus arteriosus, a normal fetal communication, facilitates the shunting of oxygenated blood from the pulmonary artery to the aorta, away from the lungs. Normally , functional closure of the ductus occurs within a few hours of birth; it is abnormal - if it persists beyond the neonatal period. The ductus closes in response to an increase in peripheral oxygen saturation and a drop in the resistance of the pulmonary circulation as the lungs expand; this causes the ductal tissue to contract through a prostaglandin inhibition mechanism. A cyclo-oxygenase inhibitor (e.g. indomethacin) may be used therapeutically to close the ductus in the first few weeks of life. In premature - babies the ductus is more likely to remain patent for longer or permanently . In the isolated case of PDA, there is a left-to-right shunt of blood, resulting in a high pulmonary blood flow . Small shunts usually cause few symptoms and signs apart from the continuous machinery murmur in the left second intercostal space. Larger ducts cause cardiac failure and can uncommonly lead to shunt reversal with cyanosis and clubbing. The diag - nosis is best confirmed by echocardiography with colour flow - Doppler imaging. After 6 months of age, PDA closure is rare. Most should - be closed by preschool age, regardless of symptoms, if the risks of infective endocarditis, left ventricular failure or , rarely , Eisenmenger syndrome are to be avoided. In the adult, surgical treatment is indicated if there is a persistent left-to-right shunt, - even in the presence of reversible pulmonary hypertension. In - the premature infant, if medical treatment to close the duc - tus is unsuccessful, the PDA may be treated by percutaneous interventional cardiology techniques using an umbrella or coil duct occlusion device. If the PDA is very large or the patient very small, surgical closur e via a left thoracotomy is preferred. This can be accomplished by either ligation or division of the PDA. The operative mortality rate is low and outcome gener - ally very good. Coarctation of the aorta - This accounts for 6–7% of congenital heart disease and is defined as a haemodynamically significant narrowing of the aorta, usually in the descending aorta just distal to the left subclavian artery , around the area of the ductus arteriosus (Figure 59.24). The coarctation typically puts a pressure load on the left ventricle, which can ultimately fail. The upper body is well perfused but the lower body , including the kidneys, is poorly perfused, leading to fluid overload, excess renin secre tion and acidosis. Coarctation usually a ff ects boys and, if it occurs in girls, is suggestive of Turner syndrome. In the neonatal period, coarctation (‘infantile’ or preductal coarctation) presents with symptoms of heart failure. The child may appear well in the first few days of life because the coarcta tion is bypassed by the ductus arteriosus and o xygenated blood reaches the systemic circulation. As the ductus closes, the child

becomes progressively more unwell. In adult-type coarctation (juxtaductal or slightly postductal) obstruction is gradual with complications dev eloping in adolescence or early adulthood. Hypertension is a common presenting problem in older chil dren – often upper body hypertension only with development of enormous collateral vessels that may cause rib-notching and flow murmurs over the scapula. Other symptoms include prominent pulsation in the neck, tired legs or intermittent claudication on exercise. Clinical examination of the pulses may demonstrate a radio-femoral delay and a murmur that is continuous and heard best over the thoracic spine or below the left clavicle. The chest radiograph classically demonstrates rib-notching because of dilated posterior intercostal vessels. The heart is usually of normal size in the older child and shows a classical ‘three sign’ replacing the typical aortic knuckle . The upper part of the three sign is the dilated left subclavian, the middle part is the narrowing at the coarctation site and the lower part is the poststenotic dilatation of the descending aorta. presents with cardiac failure, often requiring vigorous medical treatment, including the administration of prostaglandin to reopen the ductus and general resuscitation before corrective surgery . Definitive treatment is usually surgical repair via a left thoracotomy . Coarctation presenting in the child or later typically requires surgical repair, as most patients die before the age of 40 years because of the associated complications. Percutaneous stenting is currently the standard treatment for adults with isolated coarctation. Without correction, the majority of deaths are caused by heart failure, infective endocarditis, rupture of the aorta or haemorrhagic stroke. The preoperative hypertension may not resolve despite surgical repair. Atrial septal defects An ASD is a defect in the septum between the left and right atria leading to a left-to-right shunt, the significance of which is determined by the size of the defect and the relative compli - ance of the ventricles. The development of the atrial septum is complex and abnormalities of development lead to three commonly recognised ASDs (Figure 59.25). The most common type is an ostium secundum ASD. The anomaly is caused by failure of the septum prim um to develop, leading to incomplete coverage of the ostium secundum. These defects are usually asymptomatic in childhood, with symptoms developing insidiously , typically presenting in middle age with congestive cardiac failure secondary to pulmonary hyperten - sion or with atrial arrhythmias. In ostium primum ASD, the anomaly is a form of partial atrioventricular canal defect or endocardial cushion defect. The abnormalities are confined to the atrial septum and are - caused by the endocardial cushions failing to develop and so close the ostium primum part of the interatrial se ptum. The defect is associated with abnormalities of the mitral valve, lead - ing to mitral regurgitation. There is a relatively high incidence of this abnormality in trisomy 21 (Down syndrome). Typically , - -

Coarctation Subclavian artery Ligamentum arteriosum Ascending aorta Pulmonary Intercostal artery

arteries Internal thoracic artery Descending aorta Figure 59.24 Coarctation of the aorta. Coarctation causes severe obstruction of blood /f_l ow in the descending thoracic aorta. The descending aorta and its branches are perfused by collateral channels from the axillary and internal thoracic arteries through the intercostal arteries (arrows). SVC Sinus venosus or superior Tricuspid caval defect valve Fossa ovalis defect Atrioventricular defect IVC Coronary sinus Figure 59.25 Atrial septum viewed from

the right. The fossa ovalis is a useful reference point; the most common defect is in this area and is called a fossa ovalis (or ostium secundum) defect. A defect near the atrioventricular junction may be part of the spectrum of atrioventric

ular septal defects; defects near the entry of the superior vena cava (SVC) are commonly associated with anomalies of venous drainage into the atria. IVC, inferior vena cava.

hood, with dyspnoea, recurrent chest infections and, if pulmo nary hypertension develops, cyanosis. A sinus venosus ASD is a rare defect and is the result of failure of partition of the pulmonary and systemic venous cir culations. These defects are most commonly located high in the atrial septum at the junction of the superior vena cava and the right a trium. They are frequently associated with anoma lous pulmonary venous drainage, with right superior pulmo nary veins draining into the superior vena cava or right atrium directly . Summary box 59.12 Atrial septal defects (ASDs) /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF Closure is performed during the first decade of life, even in the absence of symptoms, to avoid late-onset right ventric ular failure, endocarditis and paradoxical emboli. In adults, closure is still appropriate for symptomatic improvement and avoidance of complications. The traditional method of clo sure in volves open-heart surgery with CPB and closure of the defect, either directly with sutures, as with most secundum defects, or, if the defect is large, using a pericardial or synthetic patch. Closure of small to moderate ASDs using percutaneous catheter-delivered devices in the cardiology catheter labora tory is increasingly common. Primum atrioventricular defect repairs may require additional mitral valve repair. The opera tive mortality rate for isolated atrioventricular defect repairs is <1%, with an excellent prognosis. Surgical correction of com plete atrioventricular canal defects, with closure of the ASD and ventricular septal components and mitral v alve repair, is possible, but with a higher surgical mortality rate. Ventricular septal defects A VSD is a defect in the interventricular septum that allows left-to-right shunting of blood. VSDs account for 20–30% of congenital heart

disease and a ff ect approximately 2 in 1000 live births. They may occur in isolation or as part of a more complex set of cardiac abnormalities (e.g. tetralogy of Fallot, Henri Louis Roger , 1809–1891, physician, Hôpital Sainte-Eugene, Paris, France. - Types of ventricular septal defects (VSD) - /uni25CF - - /uni25CF /uni25CF /uni25CF complete atrioventricular canal defect). Four major anatomical types of VSD are described, based on the anatomical subsec - tions of the interventricular septum (Figure 59.26). The VSD permits a ventricular left-to-right shunt, with subsequent right ventricular volume overload and increased pulmonary blood flow . This may lead to progressive pul - monar y oedema and congestive cardiac failure. Persistently elevated pulmonary blood flow and pulmonary vascular resis - tance also lead to irreversible pulmonary h ypertension. They may eventually result in reversal of flow across the defect and Eisenmenger syndrome. The clinical presentation reflects the magnitude of the left-to-right shunt, which, in turn, depends on the size of the VSD and the pulmonary and systemic vascu - - lar resistances. Small defects may close or cause little systemic disturbance (maladie de Roger); infants are asymptomatic with normal development. In the first 5 years, up to 50% of VSDs - close spontaneously . Clinically , a loud pansystolic murmur can - - -

Common defects Ostium secundum: fossa ovalis defect (approximately 70% of ASDs) Ostium primum: atrioventricular septal defect (approximately 20% of ASDs) Sinus venosus defect: often associated with anomalous pulmonary venous drainage (approximately 10% of ASDs) Patent foramen ovale: common in isolation, usually no left-to- right shunt (not strictly an ASD) Rarer defects Inferior vena cava defects: a low sinus venosus defect and may allow shunting of blood into the left atrium Coronary sinus septal defect: also known as unroofed coronary sinus, with the left superior vena cava draining to the left atrium as part of a more complex lesion Perimembranous (conoventricular) defect The most common defect (70–80%), usually located within the membranous septum and may extend to the tricuspid valve annulus or the base of the aortic valve Muscular (trabecular) defect Occurs in 10% of cases and is located within the membranous septum and can be multiple Atrioventricular (inlet) defect Also called an atrioventricular canal-type defect; occurs in 5% of cases and is located in the atrioventricular canal beneath the tricuspid valve Subarterial (outlet) defect Occurs in 5–10% of cases and lies within the conal septum immediately beneath the pulmonary valve annulus Subarterial Perimembranous Atrioventricular Muscular Figure 59.26 Ventricular septum viewed from the right, showing the characteristic sites of ventricular septal defects.

flow between the ventricles. Large defects typically present with congestive cardiac failure in the first 2 months of life. Because of the size of the VSD, ventricular pressures are equalised and often only a soft systolic murmur is detected. If left untreated, pulmonary hypertensive changes start from about 1 year of age. Eisenmenger syndrome, secondary to shunt reversal in such cases, may become evident in the second decade of life. Echocardiography confirms the diagnosis and can estimate the degree of shunting across the defect. Cardiac catheteri sation can quantify right and left cardiac pressures and the degree of pulmonary hypertension, as well as demonstra step-up in oxygen saturation between left and right ventricles. Generally , surgical closur e is indicated for large defects; when there is failure to respond to medical therapy; for left-to-right shunts of >2:1; when there are signs of increasing pulmonary vascular resistance; and in the presence of complications of VSD. These include: (i) aortic regurgitation, which occurs in about 5% of defects; (ii) infundibular stenosis, which tends to be progressive and leads to shunt reversal; and (iii) infective endocarditis, often presenting with pneumonia or pleurisy as the infected ‘emboli’ in a VSD with a typical left-to-right shunt flows into the pulmonary circulation.

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