

Classification

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Congenital heart disease can be broadly classified according to the presence or absence of cyanosis, although the distinction is not always clear-cut. Central cyanosis – blueness of the trunk and mucous membranes – results from levels of deoxygenated haemoglobin of $>3-5$ g/dL in the arterial circulation. Cyanotic congenital heart diseases make up 25% of cases (8 or 9/1000 live births) and are usually more complex, - although they do include simple defects. Cyanotic congenital cardiac lesions can involve: monary blood flow . Many lesions consist of septal defects in conjunction with a right-sided obstructive lesion, producing obligatory right-to-left shunts. The most common cause of this is the tetralogy of Fallot. Parallel systemic and pulmonary blood flow . If there is no mixing this is incompatible with life; neonates have a patent foramen ovale or VSD that allows some mixing of the two circulations at this level. The most common example of this is TGV . Defects in the connections of the heart in which there is mixing of the systemic and pulmonary flows. An example of such a complex lesion is total anomalous pulmonary venous drainage (TAPVD). Acyanotic congenital heart diseases represent 75% of cases and are usually less complex. They result in an increase in the work imposed on the heart because of either: A left-to-right shunt with increased pulmonary blood flow , causing an increase in volume work of the heart. Examples include PDA, ASD and VSD. Obstruction of blood flow across a left-sided heart valve, such as aortic stenosis, or in the aorta itself, as occurs with coarctation of the aorta, leading to an increase in pressure and work of the heart. Typically , acyanotic congenital heart disease presents as heart failure in infancy because of pulmonary congestion caused by increased pulmonary blood flow or increased pulmonary venous blood pressure resulting from an obstructive lesion. The common acyanotic cardiac defects can also present as a murmur in infancy or later. Tetralogy of Fallot - This is the most common cyanotic congenital heart disease in children surviving to 1 year and accounts for about 4-6% of all congenital heart diseases. The four intracardiac lesions - originally described (Figure 59.22) were: VSD; overriding aorta; pulmonary (infundibular or subpulmonary) stenosis; right ventricular hypertrophy . - There may be no initial clinical signs, but, as pulmonary stenosis progresses, cyanosis typically develops within the first year of life. Squatting is an adaptation by the child to hypoxic spells , increasing systemic vascular resistance and the venous return to the heart. Consequently blood is diverted into the pulmonary circulation, increasing oxygenation. Lethargy and tiredness are also common. Plain radiography classically demonstrates a 'boot-shaped' heart with poorly developed lung vasculature. The diagnosis is confirmed with echocardiography . Surgical correction is the mainstay of treatment and is usually carried out at 4-6 months of age, when possible. Repair is achieved using a patch to close the VSD and resection of the obstructing infundibular septum. Surgical results are good, with a late survival rate of 95% at 5-10 years following correction of tetralogy , an operative mortality rate for a repair of between 5% and 10% and an incidence of reoperation following tetralogy repair of 5-10%. Transposition of the great vessels This is the second most common cyanotic congenital heart

disease and most common cause of cyanosis from a congenital cardiac defect discovered in the newborn period. TGV results from abnormal development, with the aorta arising from the right ventricle and the pulmonary artery from the left ventricle (Figure 59.23). The resulting transposition causes pulmonary and systemic circulations to run in parallel rather than in series; oxygenated pulmonary venous blood returns to the lungs and desaturated systemic venous blood is pumped around the body . The situation is incompatible with life and mixing of the

1 Narrowing of the pulmonary
valve 3 Displacement of aorta over
ventricular septal defect 2
Thickening of wall of right ventricle
4 Ventricular septal defect opening
between the left and right
ventricles Figure 59.22 Fallot's
tetralogy. Four abnormalities that
result in insuf /f_i ciently
oxygenated blood being pumped
to the body. Aorta Pulmonary Right
artery atrium Left ventricle Right
ventricle

Figure 59.23 Transposition of the great vessels.

foramen ovale or VSD. Patients often present with severe central cyanosis occur ring within 48 hours of birth. However, if there is a large ASD or VSD there may be minimal cyanosis initially . Typically , progr ess is poor and, as pulmonary vascular resistance declines in the neonatal period, high pulmonary flow develops, with cardiac enlargement and left ventricular failure. The chest radiograph shows pulmonary plethora, with the heart having an 'egg on its side' appearance, with a small pedicle (aorta in front of pulmonary artery). Cardiac echocar diography is su ffi cient to confirm the diagnosis and delineate the ana tomy . Many infants will die without treatment within 1 month of birth. Initial stabilisation can be achieved by performing percutaneous balloon septostomy to increase the systemic arterial oxygen saturation. Alternatively , intravenous pros taglandins can be administered to keep the PDA open and increase systemic-pulmonary shunting. Arterial switch repair is currently the standard operation and is typically carried out within the first few weeks of life. Long-term outcomes of the operation are excellent and many patients achieve good exer cise tolerance; however, some patients will require reoperation for neopulmonary stenosis. Total anomalous pulmonary venous drainage TAPVD accounts for 1-2% of congenital heart disease. In TAPVD, the pulmonary venous drainage has disconnected from the left atrium and drains into the systemic venous circu lation at some other point (inferior vena cava, superior vena cava, coronary sinus or right atrium). TAPVD presents after the first week of life with cyanosis that is mild to moderate depending on pulmonary flo w . Infants with high pulmonary flow develop cardiac failure, recurrent chest infections, failure to thrive and feeding di ffi culties. If high pulmonary flow is associated with a large ASD, cyanosis is often minimal and the lesion is tolerated well. If there is additional venous obstruc tion, cyanosis presents at birth with dyspnoea and pulmonary oedema. Echocardiography and cardiac (pulmonary) angiog raphy are necessary to confirm the diagnosis and delineate the anomalous drainage. The surgical principle is to re-es tab lish the pulmonary venous drainage into the left atrium. The exact operative tech nique depends on the anatom y and type of TAPVD. The long ter m results for survivors of the operation are generally good. Late death following repair is uncommon but, when it occurs, it is often caused by intimal fibroplasia of the pulmonary veins awa y from the anastomosis. Eisenmenger syndrome Eisenmenger syndrome is becoming less common due to development of corrective techniques for congenital heart disease with fewer patients developing a fixed increase in their pulmonary vascular resistance. It follows reversal of a left-to right shunt, that occurs with, for example, a ASD or VSD, such Victor Eisenmenger , 1864-1932, Austrian physician who described this condition in 1897, but the term 'Eisenmenger syndrome' was introduced in 1958 by an Australian cardiologist, Paul Hamilton Wood (1907-1962). cyanosis. These congenital anomalies (ASD, VSD) cause an - increase in flow and higher right-sided pressures, which lead to compensatory right ventricular hypertroph y and a subsequent rise in pulmonary artery pressure. Increasing pulmonary hypertension leads to equalisation of pressures either side of the shunt but, at some point, the right-sided pressures will exceed those on the left side, resulting in shunt reversal and desaturated blood entering the left side of the circulation. Cyanosis and dyspnoea are the most common clinical features. - Closure of the shunt is contraindicated if pulmonary hyper - tension is irreversible because the right-to-left shunt now serves to decompress the pulmonary circulation.

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