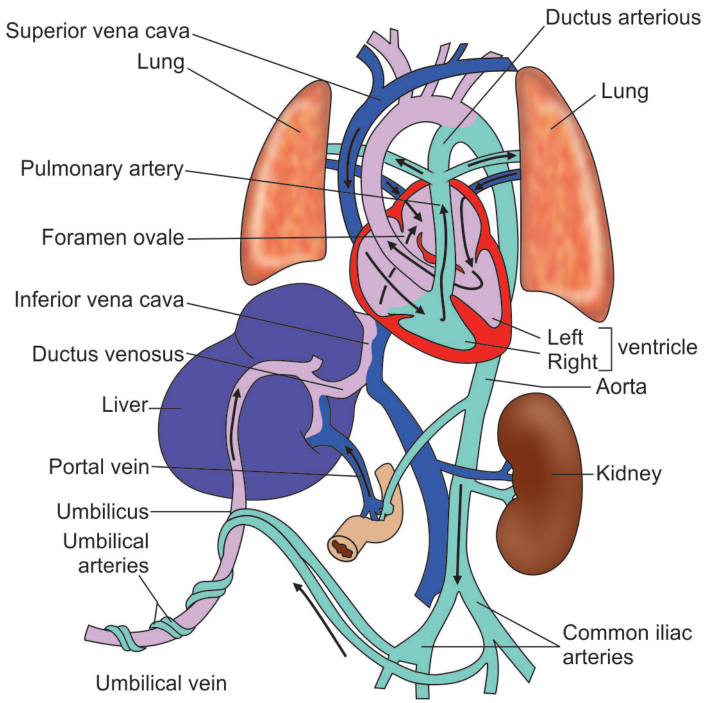

Fig. 1.5: Foetal circulation—the normal circulation of a foetus follows a slightly different path than after a baby is born

right and left atrium. It allows mixing of blood between the two atria.

In more detail, the unborn baby receives oxygenated blood and nutrients from the placenta through the umbilical vein. Most of this blood bypasses the liver through the ductus venosus and enters the inferior vena cava (the amount of blood directed towards the liver increases with increasing gestational age). Again, most of this blood returning to the heart bypasses the right ventricle and the lung circulation and flows through the patent foramen ovale (PFO) into the left atrium, left ventricle, and into the aorta. This blood, high in oxygen and nutrients, supplies the brain and upper body of the unborn child. Some of the blood that enters the right atrium via the inferior vena cava mixes with the blood returning to the right atrium via the superior vena cava, then enters the right ventricle and pulmonary artery. Most of this blood (about

Fortunately, advances in non-invasive imaging have allowed cardiac catheterization to become increasingly a catheter-based therapeutic option rather than a diagnostic tool. Echocardiography, magnetic resonance imaging (MRI), and computed tomography (CT) scan, in many cases replace the need for cardiac catheterization.

Echocardiography

Nowadays, the initial investigation of choice, in most centres, is echocardiography, which has gained particular importance in paediatric cardiac disease because of the excellent descriptions of intracardiac structures that can be obtained with high-resolution transducers. Transthoracic echocardiography is the most widely used echocardiogram in small infants.

Transoesophageal echocardiography (TEE) is done through the oesophagus with the ultrasound transducer positioned behind the heart. Transoesophageal probes are available for children as small as 3 kilograms. This technique is routinely used in the operating room to evaluate and plan procedures (Fig. 1.10).

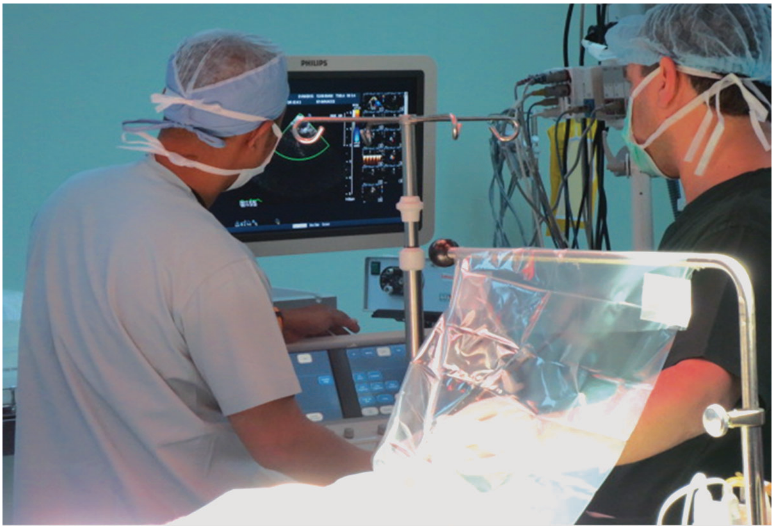

Fig. 1.10: Transesophageal echocardiography (TEE) performed in the operating room. *(Photo by EDJ, courtesy of KAMC, National Guard Hospital, Riyadh, Saudi Arabia)*

with cardiac lesions require lifelong specialized cardiac care, first with a paediatric cardiologist and later with an adult congenital cardiologist. There are more than 1.8 million adults living with congenital heart defects worldwide.

PROGNOSIS

How well a child does depends on the specific defect. Fifty years ago, the risk of death from congenital heart surgery was 30% compared to 5% today. In the 1980s, 60% of deaths from congenital heart disease occurred in the first year of life, whereas in the 1990s the majority of deaths occurred in adults over the age of twenty. It is predicted that 78% of children born with congenital heart disease will survive into adulthood. In the United Kingdom there are close to 1,50,000 adults living with congenital heart disease. Added to this number are an estimated 1,600 cases with complex or significant congenital heart lesions and many more with simple lesions entering the adult age group each year. These figures will continue to increase over time due to the ongoing development of techniques for the diagnosis and improved management of CHD. A number of large scale, long-term studies are available that predict the outcomes in some congenital heart lesions. Among the most important of these, is a report from the paediatric cardiac surgical database of 6461 children operated on, in Finland between the years 1953 and 1989 (Table 1.5).

Table 1.5: Prognosis for adults with congenital heart disease

Good	*Intermediate*	*Uncertain or poor*
Atrial septal defect	Aortic stenosis	Transposition of the great arteries
Patent ductus arteriosus	Tetralogy of Fallot	Post arterial switch procedure
Pulmonary stenosis	Transposition of the Great Arteries	Congenitally corrected transposition
Ventricular septal defect	Post-Senning/Mustard procedure	Ebstein's anomaly of tricuspid valve
Coarctation of aorta		Single ventricle physiology

Source: *Adapted from Nieminen, Jokinen, and Sairanen, 2001.*

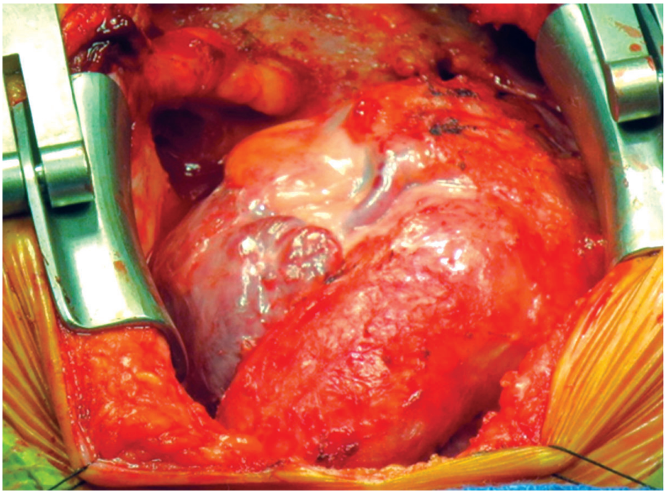

Fig. 2.4: In situs inversus, the morphologic right atrium is on the left (as seen in the picture), and the morphologic left atrium is on the right. Picture is taken standing at the head of the child. *(Photo by EDJ, courtesy of KAMC, National Guard Hospital, Riyadh, Saudi Arabia)*

Three Types of Cardiac Position

The terms levocardia, dextrocardia and mesocardia indicate only the direction of the cardiac apex.

1. **Levocardia:** The heart is located mainly in the left chest, **levo** means *left* in Latin. This is the normal position of the heart (the direction of the cardiac apex is to the left) however, some or all of the thorax or abdomen viscera are transposed laterally (situs inversus). It is also known as situs inversus with levocardia (Fig. 2.6). This condition is often associated with severe heart defects and splenic abnormalities such as asplenia (absence of normal spleen function) or polysplenia (multiple small accessory spleens). Thus in levocardia, the position of the heart is normal but other viscera such as the lungs, liver, spleen, etc. are often transposed.
2. **Dextrocardia:** The heart is located predominantly in the right chest, **dextro** means *right* in Latin (Fig. 2.7). This is abnormal (the direction of the cardiac apex is to the right). It is commonly associated with defects of the heart. Dextrocardia is believed to occur in approximately 1 in 12,000 children. Dextrocardia situs