8
Univentricular Heart (Singe Ventricle)

8.1. Unoperated Univentricular Heart

The arrangement in which both atria connect to one ventricle is described as double inlet ventricle. Univentricular atrioventricular connection by means of an absent left or right connection is frequently referred to in the literature as mitral and tricuspid atresia. In most “univentricular hearts”, there are two ventricular chambers with one major (dominant) ventricle and a second rudimentary chamber. The rudimentary ventricle is usually situated anteriorly either rightward or leftward and is more often of right ventricular morphology. The pulmonary artery usually arises posteriorly from the large main chamber, which frequently is of left ventricular morphology, whereas the aorta usually rises anteriorly from the rudimentary outlet chamber. Where the associated ventricular septal defect is small in this setting, it leads effectively to subaortic stenosis.

8.1.1. Double Inlet Ventricle

The echocardiographic hallmark of double inlet ventricle is more than 50% of both atrioventricular valves open into a single and large ventricle. This finding may be demonstrated from a variety of views, including parasternal, apical, and subcostal transducer locations (see Figures 8.1 and 8.2).

In an absent right atrioventricular connection (classical tricuspid atresia), the tricuspid atresia is usually an absent connection, but occasionally may be a valvar atresia.

In an absent left atrioventricular connection (mitral atresia), the morphology is essentially similar to that described in the classical tricuspid valve disease but involving the mitral valve. As the pulmonary venous return to the left atrium, there has to be always a communication at the atrial level for survival, although this may become restrictive with growth.
FIGURE 8.1. Apical views from two patients with univentricular hearts due to absent (A) right and (B) left atrioventricular connections.

FIGURE 8.2. Apical view showing double inlet left ventricle.