

Age/ Sex	Primary diagnosis	Previous congenital cardiac surgeries	Direct cause of death
15F	ASD	Insertion of Amplatzer device (aged 5)	No substrate identified
38F	ASD	Insertion of STAR atrial septal occluder (age unknown)	No substrate identified
25F	Ostium primum defect	None	Probable PH and history of arrhythmia
38F	Ostium primum defect with “cleft of trifoliate left AV valve	Not identified in life	Probable PH
28M	VSD of atrioventricular canal type	Not identified in life	No substrate identified
35F	Muscular inlet VSD with spontaneous closure by septal leaflet of TV	None	No substrate identified
29M	TOF	Pulmonary homograft insertion with VSD closure (aged 2), percutaneous melody valve implantation (aged 25)	Surgical related haemorrhage and ischaemia
72M	TOF	VSD closure and pulmonary valve repair (aged 50), tricuspid valve and pulmonary valve replacement (aged 69)	Biventricular fibrosis
48M	TOF with pulmonary atresia	Repair and VSD closure (aged 18)	RV fibrosis and PH
21M	TOF with pulmonary atresia	Repair, embolisation of collaterals and VSD closure (aged ~1)	RV fibrosis

23M	TOF with pulmonary atresia	Unifocalisation and left BT shunt (aged 7), right unifocalisation and right BT shunt (aged 9), radical repair with RV to PA pulmonary homograft (aged 10)	Pulmonary haemorrhage due to arteriovenous fistula
42M	TGA	BT shunt (aged 15 months), Mustard correction (aged 4), atrial baffle insertion (aged 6)	LV fibrosis
23F	TGA	Senning correction (aged 6 months)	LV fibrosis
55F	CCTGA	None	Left sided RV fibrosis
11M	CCTGA	Double switch and VSD closure (aged 11)	Right sided LV hypertrophy and left sided RV dilation
32F	CCTGA with non-compaction of LV	Pulmonary homograft, mechanical valve placement, two Amplatzer devices for paravalvular leak (ages unknown)	LV fibrosis and PH
68F	Mirror imaged atria with AV and VA concordance	None	Right sided LV fibrosis
58F	Mirror imaged atria with AV and VA discordance and VSD	None	Biventricular fibrosis, PH and peripheral PE
44F	Large perimembranous VSD with VA discordance	None	Biventricular fibrosis and PH

71F	Left atrial appendage isomerism with quasi mirroring	None	LV fibrosis and sepsis
37F	Double outlet right ventricle with double inlet left ventricle	PA banding (aged 8 months), Waterson's shunt (aged 6)	Biventricular fibrosis and PH

Table 1. The table provides the individuals age at time of death and their sex along with their primary diagnosis, previous cardiac surgeries and direct cause of death. ASD: atrial septal defect; AV: atrioventricular; BT: Blalock-Taussig; CCTGA: congenitally corrected transposition of the greater arteries, LV: left ventricle; PA: pulmonary artery; PE: pulmonary emboli; PH: pulmonary hypertension; RV: right ventricle; TGA: transposition of the greater arteries; TOF: tetralogy of Fallot; VA: ventriculo-arterial; VSD: ventricular septal defect.

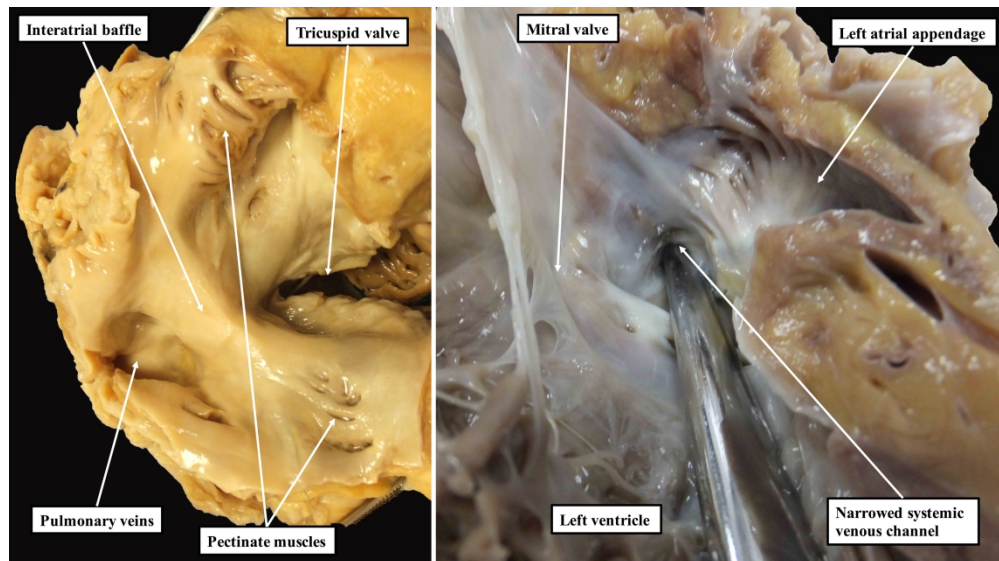


Figure 1. The images show how, in the setting of a Senning repair for so-called “transposition”, the combination of concordant atrioventricular with discordant ventriculo-arterial connections, the atrial chambers can be distinguished according to the extent of the pectinate muscles relative to the atrioventricular junctions. The left hand panel shows the pectinate muscles extending to the crux, whilst in the right hand panel they are confined to the tubular appendage. The probe is showing an obstructed systemic venous channel.

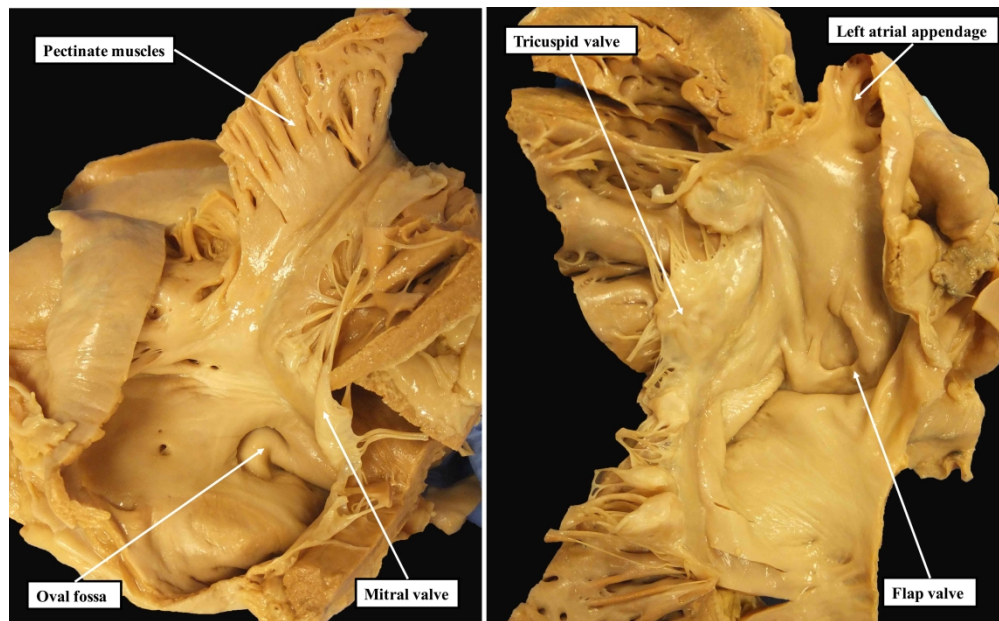


Figure 2. The images, this time from a heart with congenitally corrected transposition, the discordant ventriculo-arterial connections being “corrected” by the discordant connections across the atrioventricular junctions, show again how the atrial chambers can be recognised on the basis of the extent of the pectinate muscles, which encircle the atrioventricular junction in the left hand panel, but are confined within the appendage in the right hand panel. Note also the different features of the right and left sides of the oval fossa. The relationships of the leaflets of the atrioventricular valves to the septum show that the valve in the left hand panel is of mitral morphology, lacking tendinous cords which attach its leaflets to the septum. Such cords are seen in the right hand panel. Since the valves always insert into their appropriate ventricles, these findings indicated that the morphologically right atrium is connected to the morphologically left ventricle (left hand panel), whilst the morphologically left atrium is connected to the morphologically right ventricle (right hand panel).

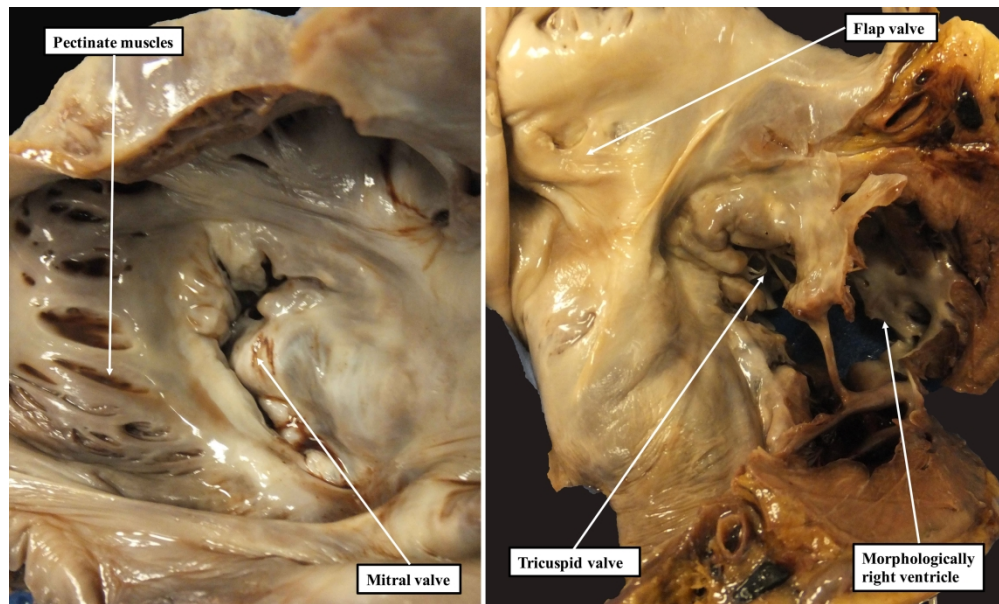


Figure 3. The images show the structure of the atrioventricular junctions in the setting of congenitally corrected transposition as found with mirror-imaged atrial arrangement. The left hand panel shows the left-sided junction. The pectinate muscles encircle the junction, showing that the left-sided atrium is morphologically right. The atrioventricular valve, however, has a solitary zone of apposition between its leaflets, identifying it as the morphologically mitral valve. This means that the left-sided ventricle is also morphologically left. The right hand panel shows the smooth vestibule of the right-sided atrium, identifying it as being morphologically left. It opens through a morphologically tricuspid valve into the right-sided morphologically right ventricle (see also Figure 4).

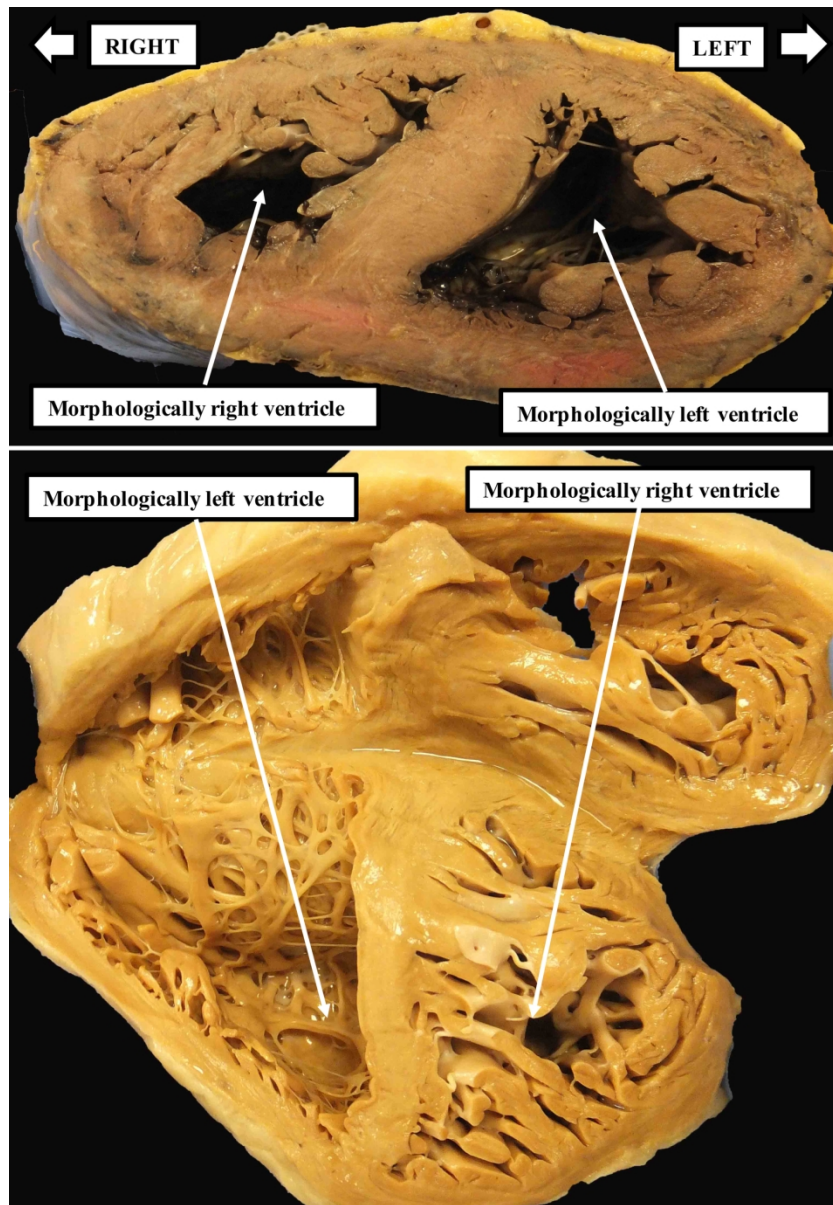


Figure 4. The images compare the arrangement of the ventricular chambers when congenitally corrected transposition is found in association with mirror-imaged atrial chambers (upper panel) as opposed to usually arranged atrial chambers (lower panel). The upper panel shows the smooth septal surface of the morphologically left ventricle, which is left-sided when associated with mirror-imaged corrected transposition (right handed ventricular topology). The lower panel shows the marked differences in apical trabecular pattern, with the morphologically left ventricle being right-sided in this example from a patient with congenitally corrected transposition and usual atrial arrangement. The ventricular mass shows left-handed topology.

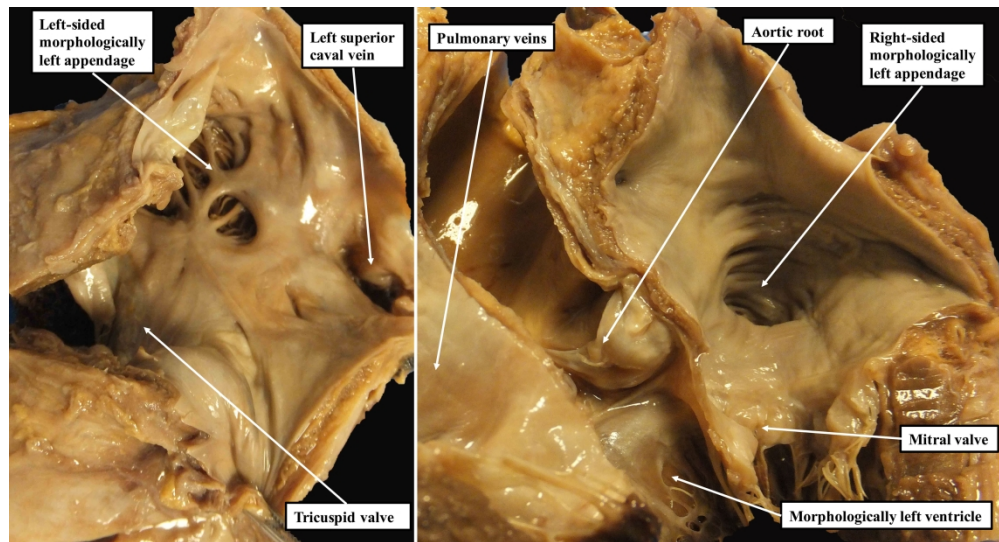


Figure 5. The images show how both atrial chambers from the heart with mirror-imaged venous returns are largely smooth-walled. The right-sided atrium (right hand panel) is connected through a mitral valve to the right-sided morphologically left ventricle, which then gives rise to the aorta, which is positioned posteriorly and to the left (mirror-imaged arterial relationships). The left-sided atrium, seen in the left hand panel, connects through a tricuspid valve to the morphologically right ventricle, which is left-sided, but which gives rise to the pulmonary trunk, which spirals in mirror-imaged fashion relative to the aortic root as it extends into the mediastinum.

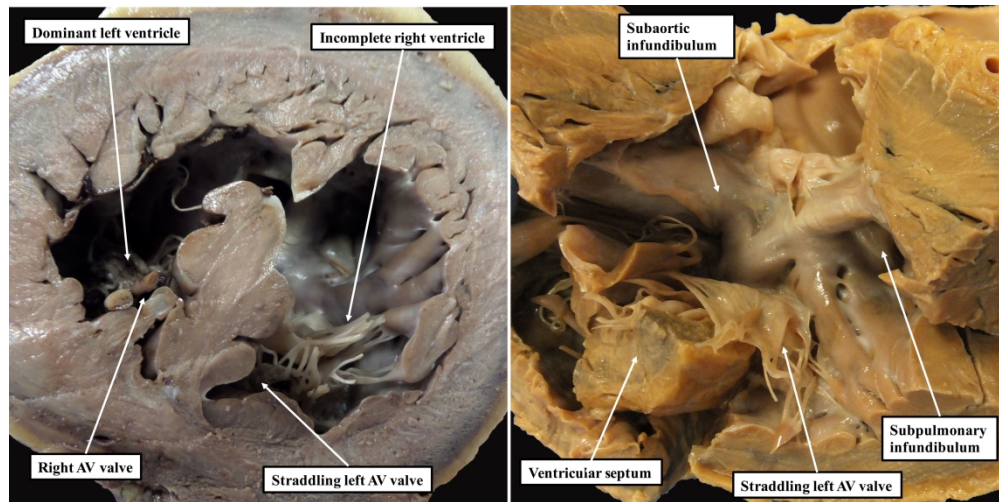


Figure 6. The images show the base of the heart before (left hand panel) and after (right hand panel) opening across the right and left atrioventricular junctions. There is straddling and overriding of the left atrioventricular (AV) valve. The atrioventricular junctions are supported mostly by the dominant right-sided left ventricle, with both arterial trunks arising from the left-sided incomplete ventricle. Both arterial valves are supported by complete muscular infundibulums. There was dome-shaped pulmonary stenosis, which had protected the pulmonary vasculature.

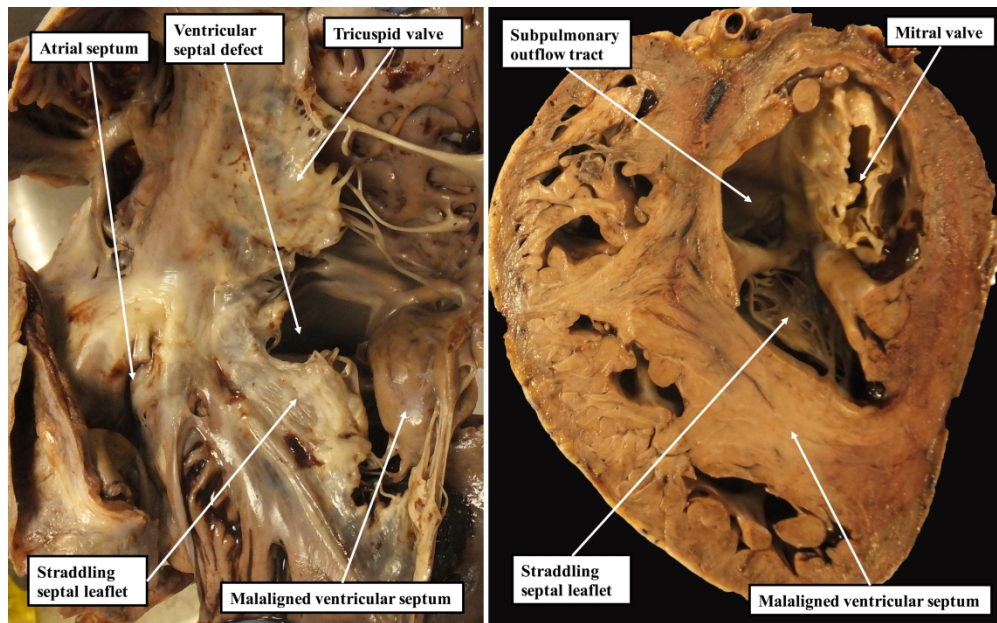


Figure 7. The images show the features of straddling tricuspid valve, but in this heart there are concordant atrioventricular connections, with discordant ventriculo-arterial connections, in other words "regular" transposition. The left hand panel shows how the tricuspid valve straddles the inferior part of the malaligned ventricular septum, which no longer extends to the crux of the heart. The right hand panel shows the base of the heart viewed from the apical aspect. The straddling septal leaflet of the tricuspid valve can be seen, along with the mitral valve, which guards a separate left atrioventricular junction. The heart does not have an "atrioventricular canal" defect.

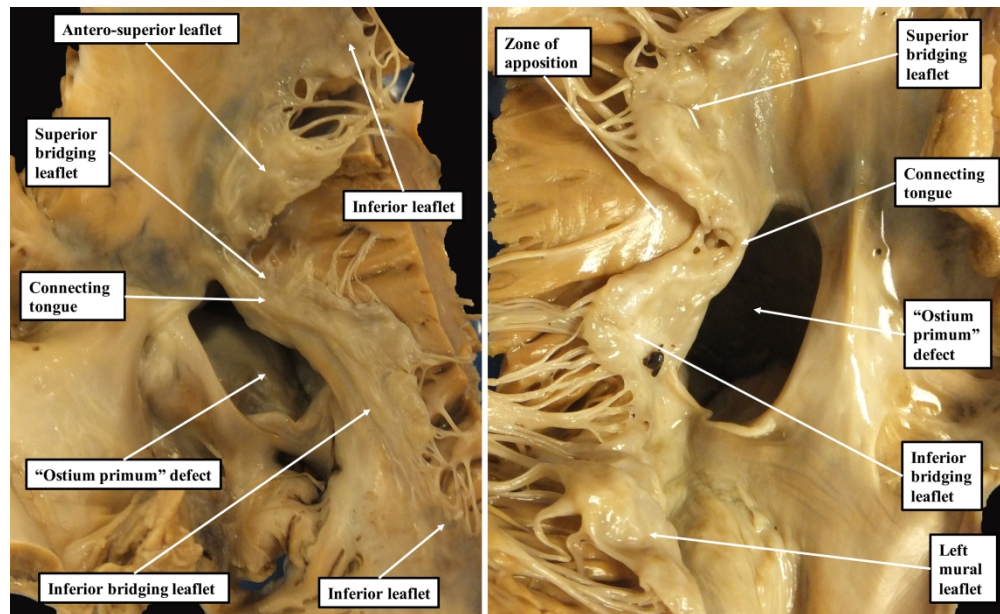


Figure 8. The images show the features of the so-called "ostium primum defect, in reality an atrioventricular septal defect in a heart with common atrioventricular junction, but with shunting through the septal defect confined at atrial level. The left hand panel shows the view from the right side. There is a tongue of valvar tissue joining the superior and inferior leaflets, creating dual orifices within the common atrioventricular junction. The right hand panel, showing the left-sided aspect, then shows how the bridging leaflets of the common valve, along with the connecting tongue, are bound down to the crest of the "scooped-out" ventricular septum. The so-called "cleft" is, in reality, the zone of apposition between the left ventricular components of the two leaflets that bridge the ventricular septum.

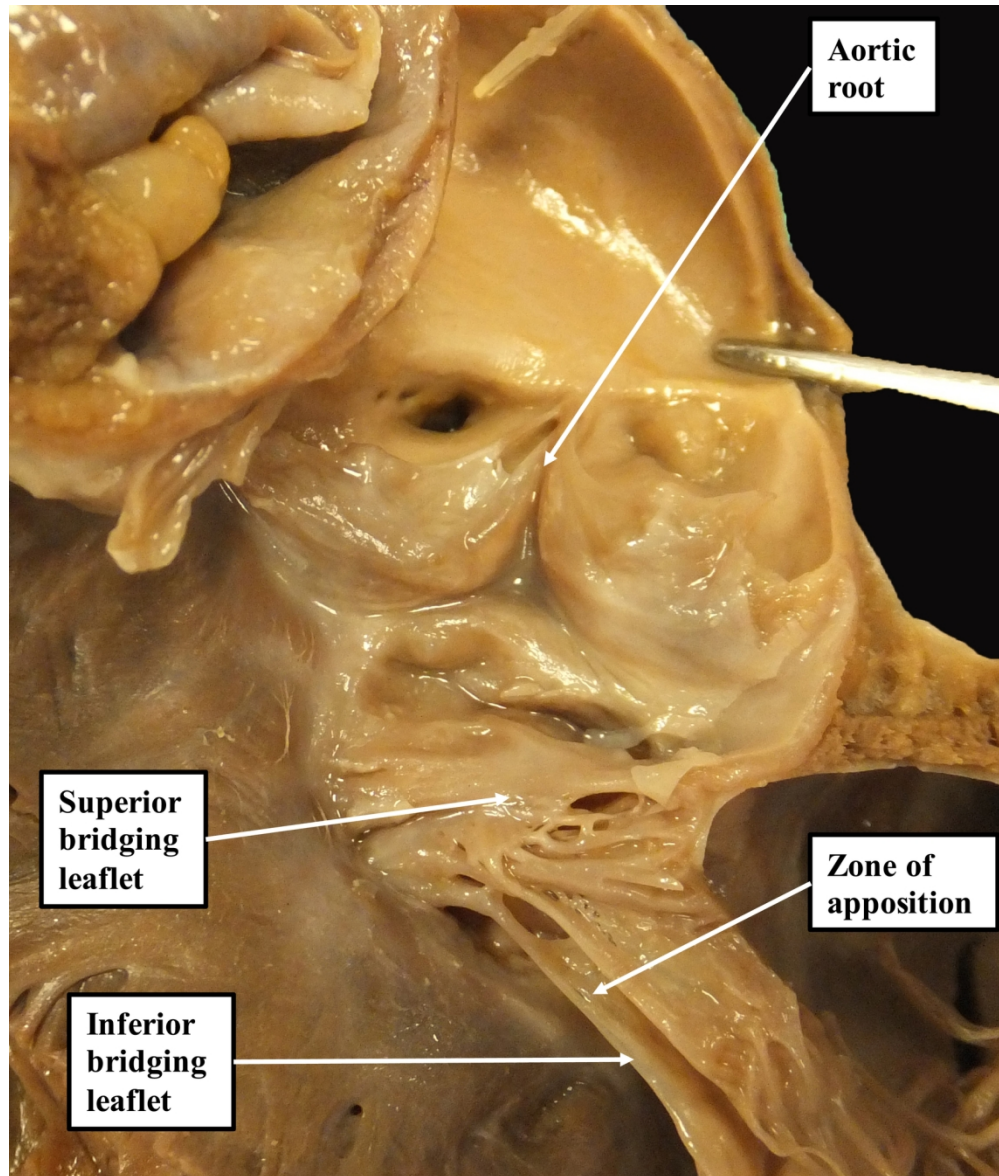


Figure 9. The image shows the left ventricular outflow tract from a heart initially diagnosed as showing a "Gerbode" defect, which is an atrioventricular septal defect in the setting of separate atrioventricular junctions. The heart, however, shows at the features of a heart with common atrioventricular junction. The aortic root is "unwedged", and the left atrioventricular valve is trifoliate.

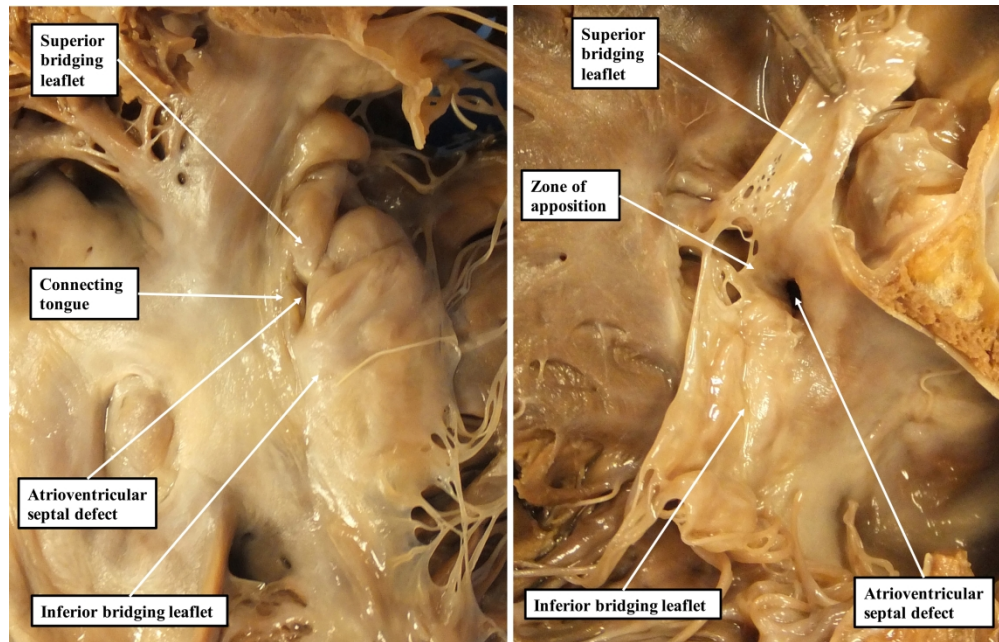


Figure 10. The images show the features of the septal defect in the heart also shown in Figure 9. As can be seen from the left hand panel, the defect opens on the ventricular aspect of the atrioventricular junction. The right hand panel shows that the tongue of tissue connecting the two bridging leaflets is attached to the leading edge of the atrial septum, thus confining shunting through the defect, which is an atrioventricular septal defect, at ventricular level.