

## Continuing Medical Education

# The aorto-ventricular tunnels

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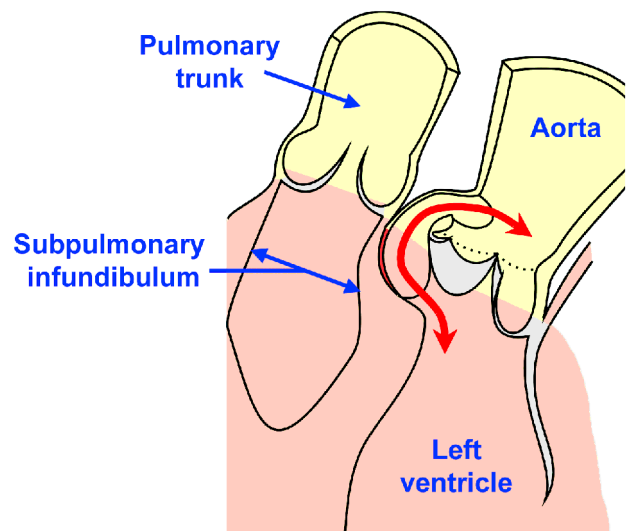
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IT IS LEVY AND COLLEAGUES, IN 1963, WHO ARE generally credited with the first description of “aortico-left ventricular tunnel”.<sup>1</sup> Examples of the malformation, nonetheless, were illustrated initially by Burchell and Edwards in 1957,<sup>2</sup> and by Edwards in 1961.<sup>3</sup> The subsequent documentation of more than 130 cases has now elucidated many features of the so-called “tunnels,” including their clinical presentation and surgical management. While most of the abnormal channels extend between the aorta and the left ventricle,<sup>4–79</sup> it is now also recognized that some, alternatively, enter the right ventricle.<sup>80–91</sup> The anatomic arrangement underscoring the malformations has been clarified by recent morphological studies,<sup>92,93</sup> while diagnosis during fetal life has established beyond any doubt that the lesions are congenital.<sup>42,43</sup> Although rare, the aorto-ventricular tunnel is the foremost cause during infancy of regurgitant flow of blood from the aorta to one or the other of the ventricles. In this review, we will describe and illustrate the structure of the malformations, speculate upon their developmental basis, and discuss pertinent aspects of their diagnosis and treatment.

### Pathologic anatomy

The aorto-ventricular tunnel is an abnormal channel that connects the lumen of the ascending aorta to the cavity of either the left or right ventricle. In its course, the tunnel forms a conduit that by-passes the sinutubular junction, this being the discrete ring

that marks the junction of the aortic valvar sinuses with the tubular ascending aorta. At the same time, the tunnel by-passes the attachment of one of the leaflets of the aortic valve, which is tethered distally at the sinutubular junction. The abnormal pathway runs into the extra-cardiac tissues as it passes from its aortic origin to its ventricular termination. In the majority of cases, these extra-cardiac tissues are those that, normally, separate the subpulmonary infundibulum from the aorta (Fig. 1). It is the origin of the tunnel



**Figure 1.**

Diagram showing the essential anatomy of a tunnel by-passing the hinge of the aortic valve to produce an aorto left ventricular tunnel. The arterial roots have been transected to show the left ventricular end of the tunnel (red arrow) as a space within the intercoronary interleaflet triangle. The tunnel runs on top of the sub-pulmonary infundibulum and into the tissue plane between the arterial trunks. It then turns rightward to exit into the ascending aorta above the sinutubular junction (dotted line) of the right coronary sinus. This leaves a bar of arterial tissue which supports part of the right coronary leaflet.

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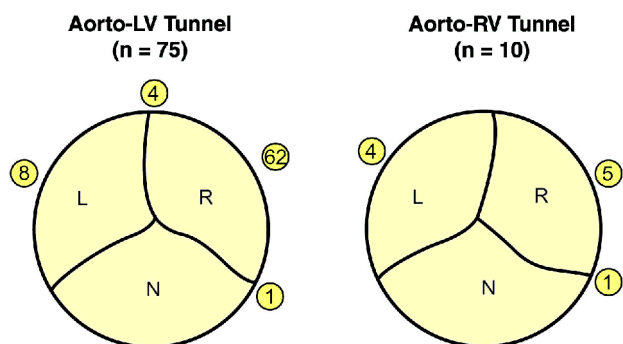
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from the tubular aorta that serves to differentiate it from rupture of an aneurysmal sinus of Valsalva. The latter lesion also creates a communication between the aorta and a ventricular chamber, but one that originates below the sinutubular junction, and remains completely within the heart. The distinction from coronary-cameral fistula is less clear, because a coronary arterial orifice may arise above the sinutubular junction, and because the left,<sup>21,84</sup> circumflex,<sup>85</sup> anterior interventricular<sup>78,91</sup> and right<sup>1,21,31,42,43,47,81,85</sup> coronary arteries have all been found arising within an aorto-ventricular tunnel. Fistulous connections of the coronary arteries, however, always pass through myocardium to reach the lumen of a cardiac chamber, and do not involve the hinge-point of an aortic valvar leaflet. As we will see, these features do not always serve to distinguish a fistula from a tunnel extending to open within the right ventricle, but they do contrast with most tunnels that open within the left ventricle. The frequent association of the tunnels with abnormalities of the coronary arteries, moreover, does suggest the possibility of a shared developmental origin.

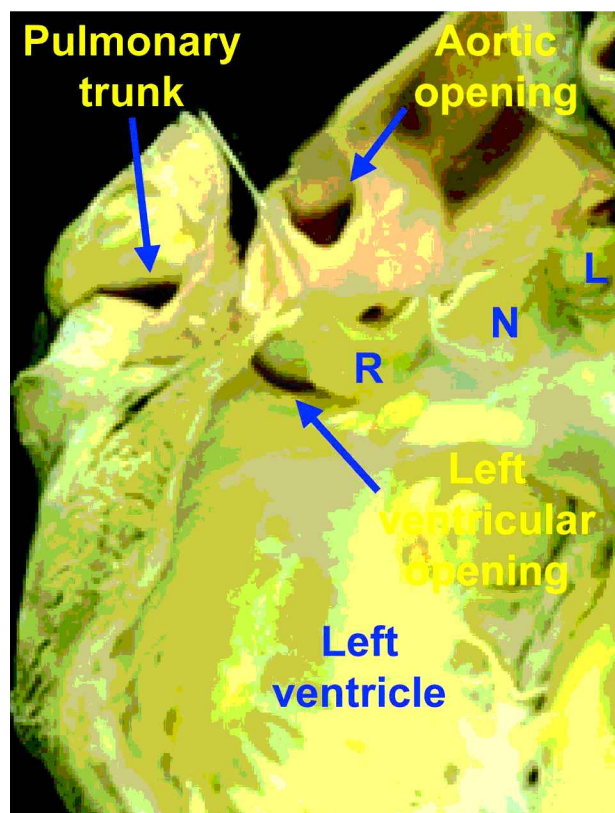
When considering the morphology of the tunnels, we will describe an aortic origin and a ventricular termination. Since the pressures will be comparable throughout the length of the tunnels, and because flow may occur in either direction, this distinction is arbitrary. In our opinion, nonetheless, it helps in accounting for the anatomic features, which can otherwise be difficult to understand. The aortic origin of a tunnel that extends to open within the left ventricle may be situated anywhere above the left or right aortic sinuses, or above the junction between the two coronary aortic sinuses. In four-fifths of reported cases, the aortic end of the tunnel is positioned above the right coronary sinus (Fig. 2). In the much smaller number of hearts in which the tunnel opens into the



**Figure 2.**  
Site of aortic opening in relation to the aortic valvar sinuses of Valsalva in 85 cases of aorto-ventricular tunnel. Circled numbers indicate the number of orifices found in each position. Abbreviations: LV: left ventricle; RV: right ventricle; L: Left coronary aortic sinus; R: right coronary aortic sinus; N: non-coronary aortic sinus.

right ventricle, more than one-third are described with the aortic origin above the left sinus of Valsalva. In some cases, clockwise or counter-clockwise rotation of the aortic root relative to its supporting ventricular attachments has been observed.<sup>6-8</sup> A single case has been described in which the aortic orifice was positioned above the junction between the right and non-coronary aortic sinuses.<sup>87</sup>

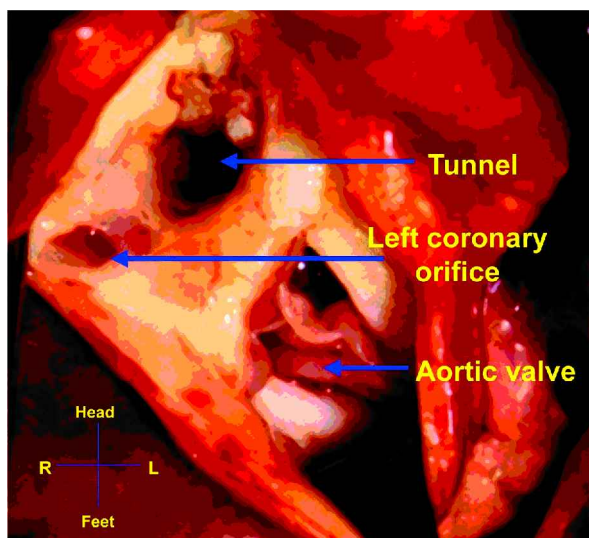
The size and shape of the aortic origin of the tunnel are extremely variable. In some hearts, the opening is a slit of only two to three millimeters width (Fig. 3).<sup>72,80</sup> At the other extreme, an oval orifice has been encountered measuring two by two-and-a-half centimeters (Fig. 4).<sup>18,44,45</sup> The size of the aortic opening shows no correlation with either the age or the size of the patient. By definition, the orifice lies above the sinutubular ridge which, itself, may be situated abnormally low,<sup>85</sup> and which can be considerably thickened.<sup>1,5,31,40</sup> Diffuse dilation of the entire ascending aorta, with further localized enlargement around the entrance of the tunnel,<sup>17</sup> may obscure the exact position of the orifice, particularly in relation to the origin of a coronary artery.<sup>17,40</sup> Aneurysms of any of the three aortic sinuses may coexist with an



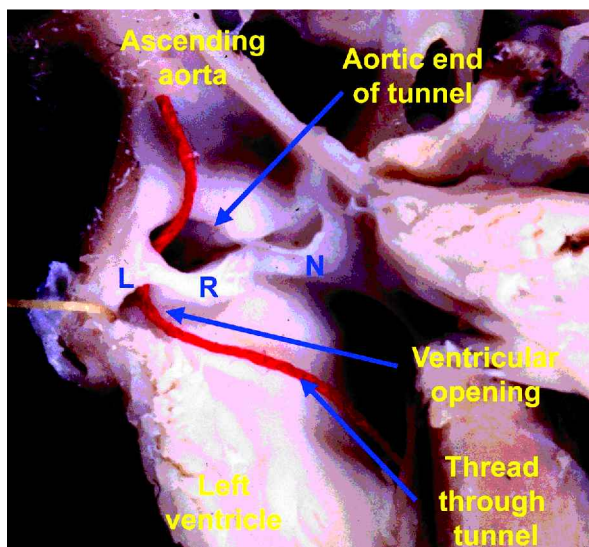
**Figure 3.**  
This tunnel has a small slit-like opening within the left ventricle, and a much larger aortic orifice which is separate from the origin of the right coronary artery. Reproduced by kind permission of Siew Yen Ho and Gaetano Thiene.

aorto-ventricular tunnel<sup>17,85</sup> Being situated below the sinutubular ridge, however, such aneurysms are distinct from the orifice of the tunnel.

Having taken origin from the aorta, the initial course of the tunnel is within extra-cardiac tissues

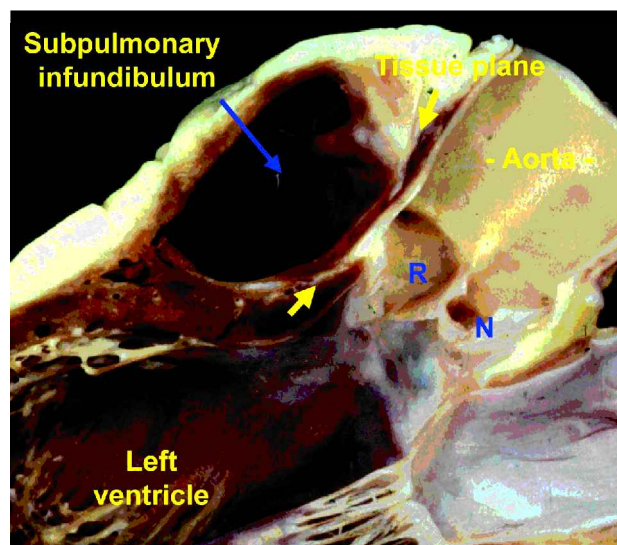


**Figure 4.** The large aortic opening of a tunnel originating above the left coronary sinus of Valsalva that extended to open in the left ventricle as seen by the surgeon at operation. Reproduced from Litwin SB, *Color Atlas of Congenital Heart Surgery*. Mosby, St Louis 1996 with permission from Mosby – Yearbook.

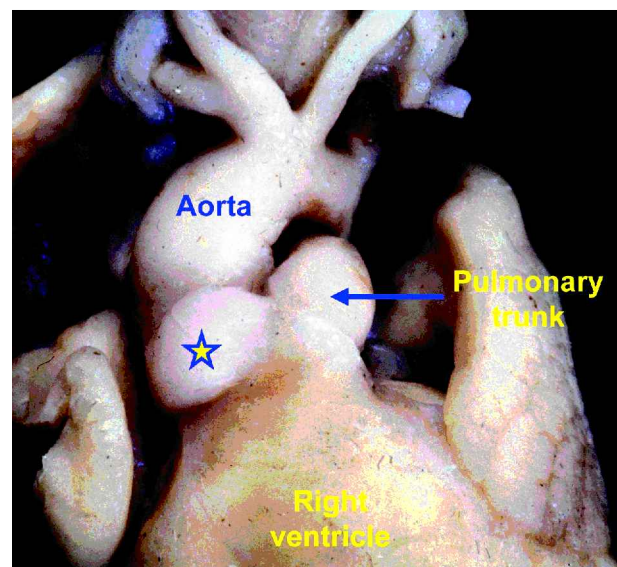


**Figure 5.** This tunnel, extending from above the right coronary aortic sinus to open within the fibrous triangle between the left and right coronary leaflets of the aortic valve, runs within the tissue plane that separates the sinuses of the aortic valve from the free-standing muscular subpulmonary infundibulum (See Fig. 7). Abbreviations: R: right coronary aortic leaflet; N: non-coronary aortic leaflet; L: left coronary aortic leaflet.

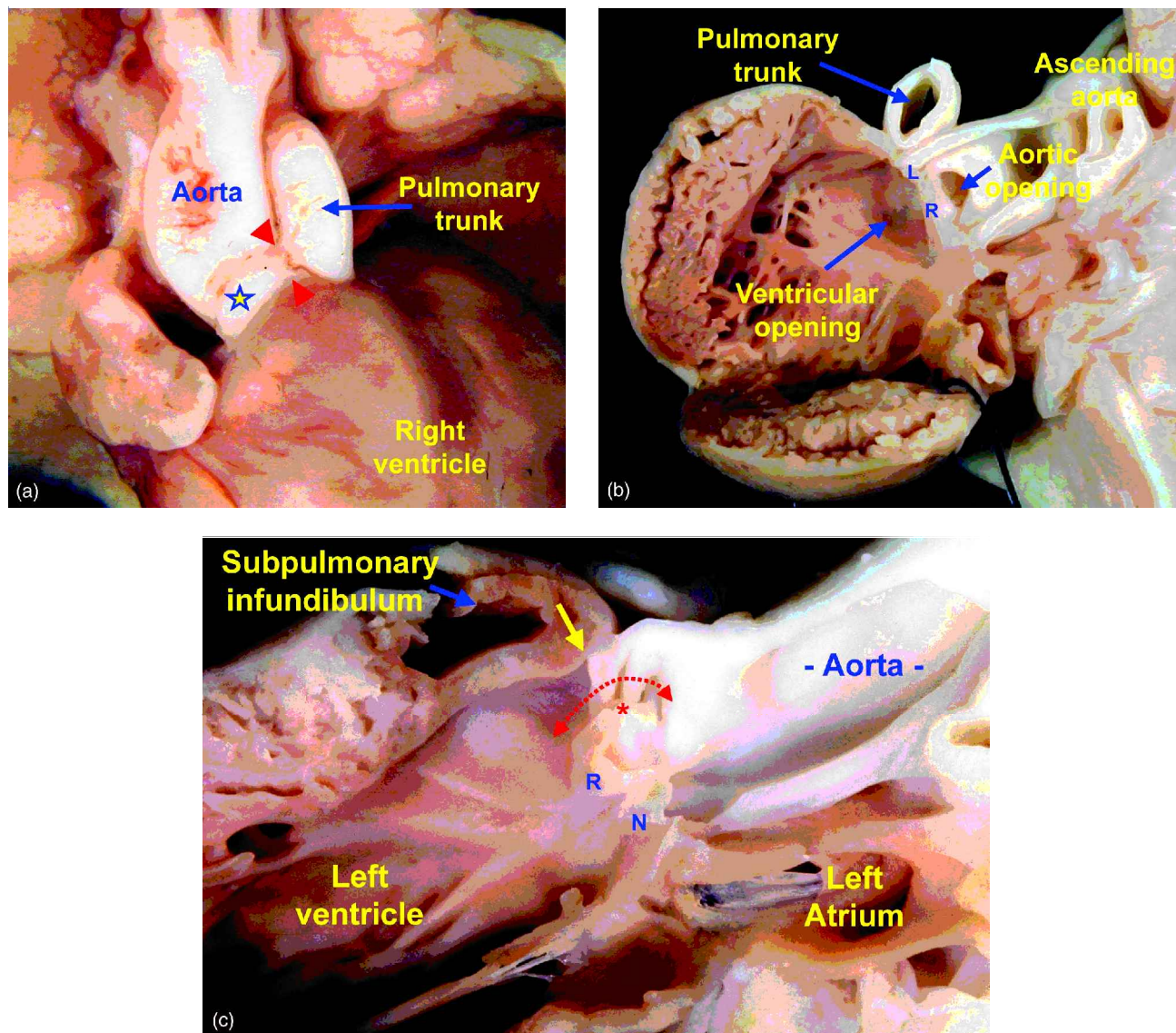
(Fig. 5), specifically in the area that, in the normal heart, forms a discrete plane between the aortic sinuses and the muscular subpulmonary infundibulum (Fig. 6).<sup>92</sup> Those tunnels that originate above the right coronary aortic sinus produce a large tubular, or saccular, protuberance on the anterior aspect of the aortic root (Figs 7 and 8).<sup>1,11,19,30,32</sup> Tunnels having their aortic end above the left coronary aortic



**Figure 6.** Sectioning the normal heart in simulated parasternal long axis plane shows the extracardiac tissue plane that normally separates the sinuses of the aorta which give rise to the coronary arteries from the free-standing muscular subpulmonary infundibulum. Abbreviations: R: right coronary aortic sinus; N: non-coronary aortic sinus.



**Figure 7.** The heart shown is the same as illustrated in Figure 5. Note the bulge made by the tunnel (starred) between the aorta and the pulmonary trunk.

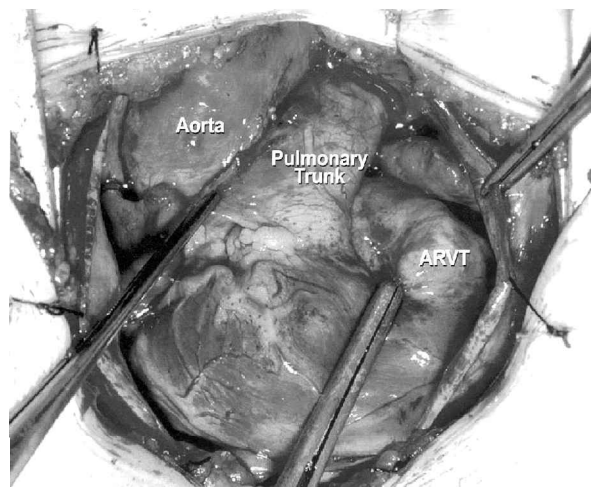


**Figure 8.**

Appearances of our most recent example of an aorto-left ventricular tunnel diagnosed at 17 weeks gestation. Examination of the arterial roots from the front (a) shows a small dilation in the region of the right aortic sinus, which is the aortic end of the tunnel (starred), as well as a prominent connection, or "arterial ligament", connecting the wall of the tunnel with the right facing sinus of the pulmonary trunk (between arrowheads). There was marked cardiomegaly, and left ventricular hypertrophy present (b). The ventricular opening of the tunnel was large, and located between the coronary aortic leaflets in the place occupied, in the normal heart, by the interleaflet fibrous triangle, which is distal to the normal anatomic ventriculo-arterial junction. In this abnormal heart, the ventriculo-arterial junction is part of the wall of the tunnel. Thus, the parasternal long axis section, taken in the same heart (c), shows that the junction between arterial wall and ventricular myocardium is still present (arrow), but has become detached from the hinge-point of the aortic valve (\*). The junction is displaced towards the subpulmonary infundibulum, and forms the mid-part of the tunnel. The tunnel itself (red arrow), therefore, bypasses the attachment of the aortic valve and enters above aortic root above the level of the sinotubular junction. Abbreviations: L: left coronary aortic leaflet; R: right coronary aortic leaflet; N: non-coronary aortic leaflet.

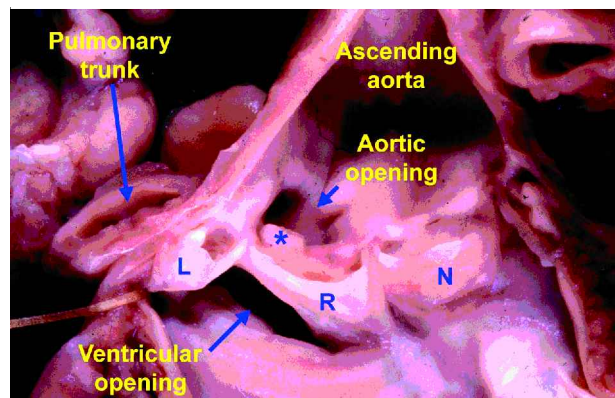
sinus, in contrast, lie behind the pulmonary trunk, originating within the transverse sinus of the pericardium.<sup>78,90,91</sup> Unless they open into the right ventricle (Fig. 9), tunnels originating above the left coronary aortic sinus are less obvious when viewed externally from the front of the heart (Fig. 10).<sup>20,44</sup> As it leaves the base of the heart, the subpulmonary

infundibulum, together with the pulmonary trunk, spirals round the right and left coronary aortic sinuses of Valsalva.<sup>93</sup> The infundibulum, therefore, is readily displaced anteriorly by a tunnel that terminates in the left ventricle, with such an arrangement potentially producing subvalvar obstruction of the right ventricular outflow tract.<sup>2,9,16,26,36,40</sup>



**Figure 9.**

The external location of a tunnel which extended from the aorta to open into the right ventricle is shown as seen in the operating room. Abbreviation: ARVT: aorto-right ventricular tunnel. Reproduced from Hruda J, Hazekamp MG, Sobotka-Plojhar MA, Ottenkamp. Repair of aorto-right ventricular tunnel with pulmonary steosis and an anomalous origin of the left coronary artery. *Eur J Cardiothorac Surg* 2002; 21: 1123–1125, with permission from Elsevier Science.

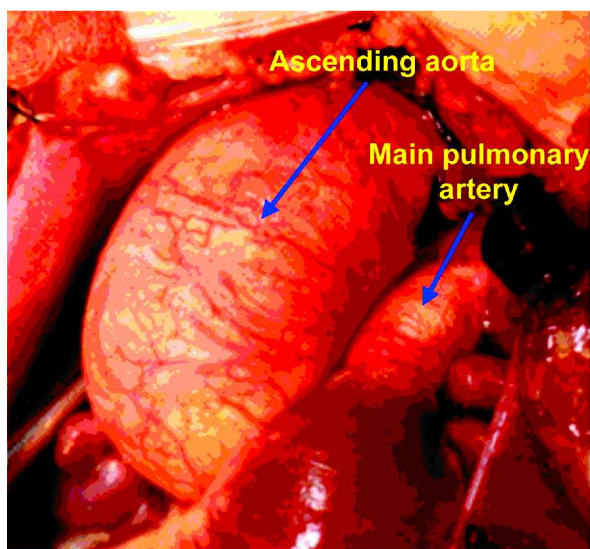


**Figure 11.**

In this heart, the left ventricle has been opened to show the termination in the fibrous triangle between the right and left coronary aortic leaflets of a tunnel that originated above the right aortic sinus. Note that the opening, although below the hinge of the valvar leaflet, is distal to the anatomic ventriculo-arterial junction. Note also the mucoid tissue in the aortic origin of the tunnel (\*). Abbreviations: R: right coronary aortic leaflet; N: non-coronary aortic leaflet; L: left coronary aortic leaflet.

to fuse with each other at the sinutubular junction (Fig. 11).<sup>92</sup> Although proximal to the sinutubular junction, this area is distal to the anatomic junction between the left ventricular myocardium and the arterial walls of the aortic root. Many tunnels have been described in surgical reports as being “immediately below the valve”. It is almost certainly the case that these open also within this interleaflet fibrous triangle. A large orifice can extend to a variable extent under the hinge of the right coronary aortic leaflet (Fig. 11).<sup>11,42</sup> Indeed, in the most recent tunnel we have investigated, the essence of the lesion was the divorce of the attachment of the right coronary aortic leaflet from its normal location within the aortic root (Fig. 8). The tunnels never extend, however, to open within the interleaflet triangle between the right and noncoronary sinuses. Thus, tunnels opening to the left ventricle are related neither to the membranous septum nor the ventricular conduction tissues.<sup>94</sup> Possible exceptions to these generalizations are rare. One is a tunnel that was said to have two adjacent ventricular orifices separated by fibrous tissue.<sup>19</sup> Another may be a tunnel noted as entering the left ventricle about ten millimeters below the aortic valve.<sup>36</sup> In a third, with associated aortic stenosis, the left ventricular communication was described as being one centimeter below the right aortic leaflet.<sup>32</sup>

Less commonly, a tunnel terminating in the left ventricle originates from the aorta above the left sinus of Valsalva. These tunnels have a much less consistent site of entry into the left ventricle. None have been documented as opening within the fibrous triangle separating the two coronary aortic valvar leaflets. Two



**Figure 10.**

As shown in this illustration, the extracardiac course of the tunnel already seen in Figure 4, which arises above the left coronary sinus of Valsalva, lies in the transverse sinus. Because of this, it is not obvious when viewed from the front of the heart. Reproduced from Litwan SB, *Color Atlas of Congenital Heart Surgery*. Mosby, St Louis 1996, with permission from Mosby – Yearbook

Those tunnels which originate above the right coronary aortic sinus, and which communicate with the left ventricle, almost always do so within the triangular fibrous area demarcated by the hinge-points of the right and left coronary leaflets as they ascend

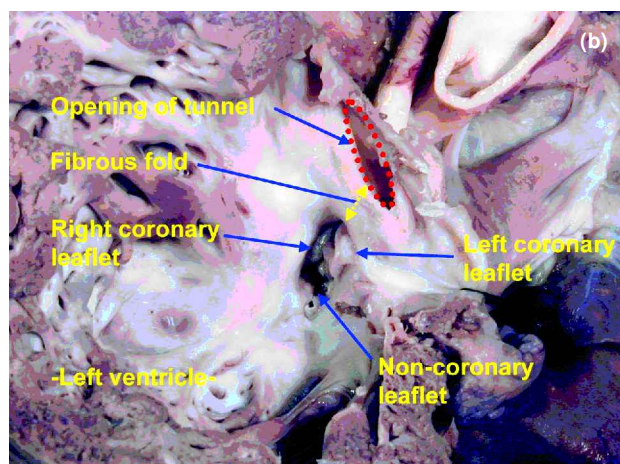
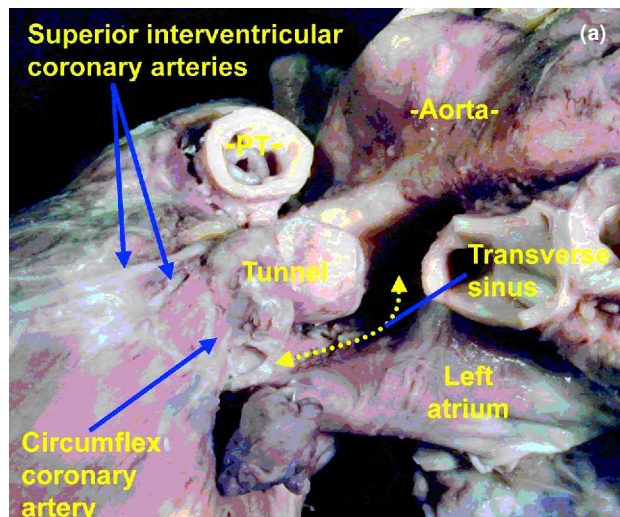


Figure 12.

The tunnel in this heart extended from above the left coronary aortic sinus (a) to open within the left ventricle proximal to the inter-coronary interleaflet triangle (b). Abbreviation: PT: pulmonary trunk.

cases have been described as opening below, and several millimeters from, the left coronary aortic leaflet,<sup>20,78</sup> another was described as opening within the crest of the muscular septum,<sup>28</sup> and still another, into the "body" of the left ventricle.<sup>13</sup> The one case within our archive had its opening below a fibrous fold, half a centimetre beneath the intercoronary interleaflet triangle (Fig. 12).

When the tunnels extend from the aorta to open within the right ventricle, the location of the ventricular orifice again shows some correlation with the position of the aortic opening. Those taking origin above the right aortic sinus of Valsalva enter the right ventricular infundibulum just proximal to the hinge-point of the pulmonary valve,<sup>84,86,87</sup> or else open within the supraventricular crest below the subpulmonary infundibulum (Fig. 13).<sup>80,83,85</sup> The ventricular orifice

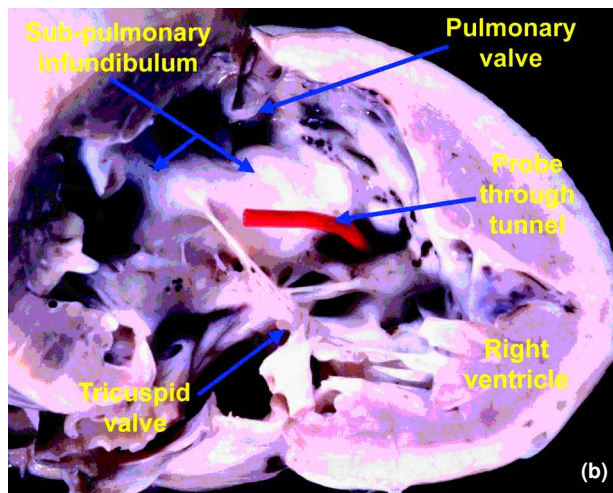
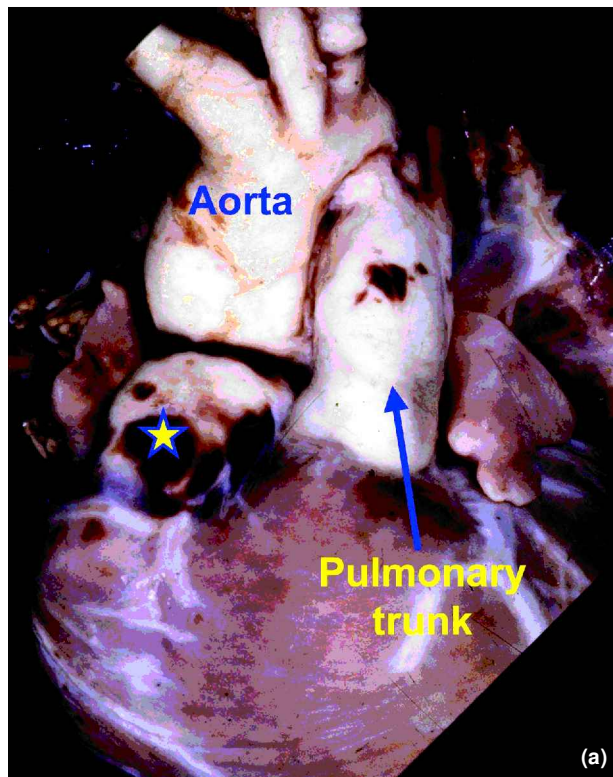


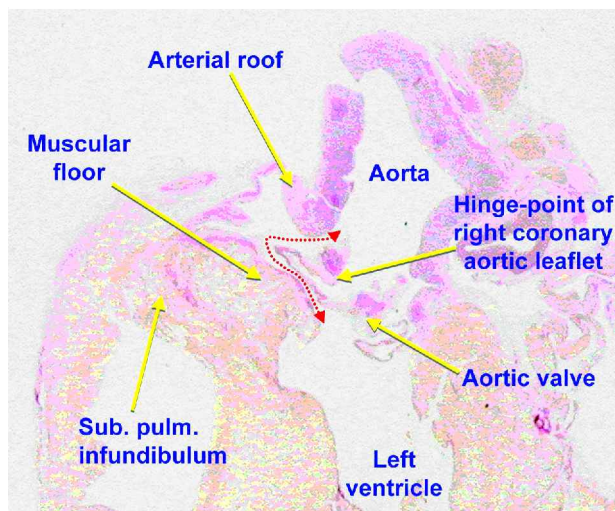
Figure 13.

A tunnel (starred) is shown that extends from the aorta above the right coronary aortic sinus (a) and opens into the right ventricle at the base of the subpulmonary infundibulum (b). Originally diagnosed as a coronary arterial fistula draining to the right ventricle, on re-examination we believe that the structure is an aorto-right ventricular tunnel.

of those tunnels which take their origin from the aorta above its left coronary sinus is more variable. Single cases have been described entering the right ventricle just below the pulmonary valvar leaflets,<sup>88</sup> in the subpulmonary infundibulum,<sup>84</sup> and in the body of the ventricle.<sup>89</sup>

Brief reflection upon the interrelations of the right and left ventricular outflow tracts<sup>92-95</sup> will confirm that, irrespective of conventional wisdom, it is exceedingly rare for the tunnels to pass through the true septal structures. This is because, in the normal heart, the interleaflet triangle separating the diverging hinge-points of the right and left coronary aortic valvar leaflets lies distal to the anatomic ventriculo-arterial junction, despite the fact that it is below the haemodynamic ventriculo-arterial junction. As already emphasized, it is through this triangular area that most tunnels empty into the cavity of the left ventricle. As a consequence of abnormal development, the crest of the muscular septum then forms the wall of the proximal part of the tunnel, being continuous with the freestanding subpulmonary infundibulum of the right ventricle (Fig. 8). Those tunnels that open into the outflow of the right ventricle pass through muscle to reach the ventricular cavity, but it is muscle that is part of the free-standing infundibulum, and not part of the muscular ventricular septum. Even tunnels which appear to lie several millimeters below the body of an aortic valvar leaflet have been found, on careful dissection, to enter the left ventricle through an in-folding of fibrous tissue, rather than passing through the left ventricular myocardium.<sup>78</sup> It is only those which end in the body of the ventricle<sup>28,89</sup> that may possibly transverse septal structures.

Histologically, the wall of the tunnel differs at its two ends. The aortic origin consists of fibrous tissue, with smooth muscle cells and elastic fibers.<sup>1,11,92</sup> This arrangement gives way to non-specific hyalinized collagen or musculature towards the ventricular opening. In reality, the "walls" of the tunnel incorporate the cardiac structures between which it passes. Thus, the usual aorto-left ventricular tunnel is confined by the musculature of the ventricular septum and subpulmonary infundibulum in its floor, and by the fibrous wall of the aortic sinus giving rise to right coronary aortic leaflet at its roof (Figs 8 and 14).<sup>92</sup> The tunnel by-passes the semilunar hinge of the right coronary aortic leaflet, which can lose completely its usual attachments to the aortic sinus and the supporting left ventricular musculature (Figs 8 and 11). Commonly, the histologic structure of the mid-portion of the tunnel is indistinct, but there can also be a clearly demarcated ventriculo-arterial junction within the body of the tunnel, lending further support to the notion that the abnormality represents separation between the attachment of the leaflet and the wall of the arterial valvar sinus (Fig. 8). Membranous or cystic structures similar to tissues of the valvar leaflets have been found within the lumen of the tunnel.<sup>15,16,72,84,85</sup> Such structures can alternatively be attached to the sinutubular ridge,<sup>31,42,78</sup> or lie



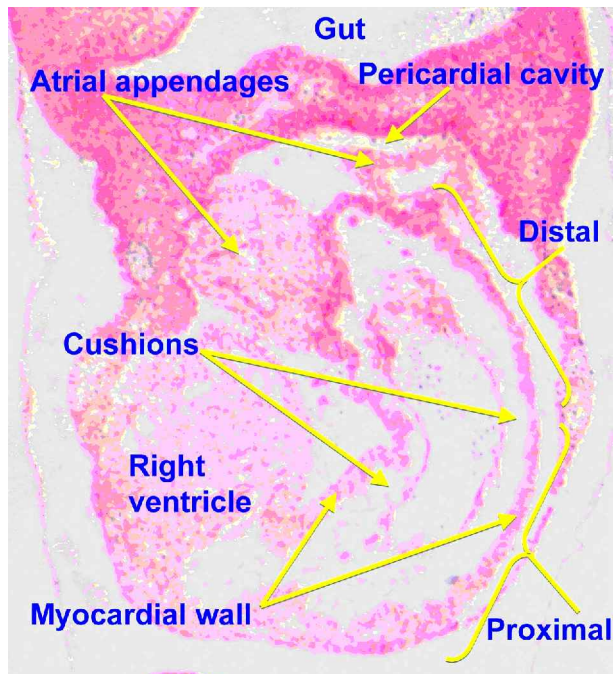
**Figure 14.** *Histological section showing how the tunnel rests on the musculature of the subpulmonary infundibulum, but that its aortic origin is of arterial phenotype.*

within an aortic sinus adjacent to the orifice of the tunnel (Fig. 11).<sup>20</sup> They can produce obstruction within the tunnel.<sup>72</sup>

Aorto-ventricular tunnels have important relationships to the proximal portions of the coronary arteries.<sup>96</sup> The orifice of the right coronary artery has been found above,<sup>19,21,40</sup> below,<sup>1,42</sup> and to the side<sup>86</sup> of tunnels situated above the right sinus of Valsalva, as well as within the tunnel itself.<sup>1,21,31,42,43,47,81,85</sup> Absence of the orifice of the right coronary artery<sup>6,21,31,41,49,87</sup> has been observed and, in one heart,<sup>85</sup> the circumflex coronary artery arose from the tunnel. When the tunnel originates from the aorta above the left sinus of Valsalva, in more than half of the reported cases an abnormal orifice of the left coronary artery has been observed to be above the tunnel,<sup>13,28</sup> within the tunnel,<sup>21,84</sup> or else to be absent.<sup>78</sup> Origin of a stenotic anterior interventricular branch from such a tunnel has also been observed.<sup>78,91</sup> A left coronary artery originating to the right of the tunnel may have an intramural course within the posterior wall of the tunnel.<sup>44</sup> Occlusion of a coronary artery by the tunnel has also been described.<sup>2,3</sup>

## Developmental considerations

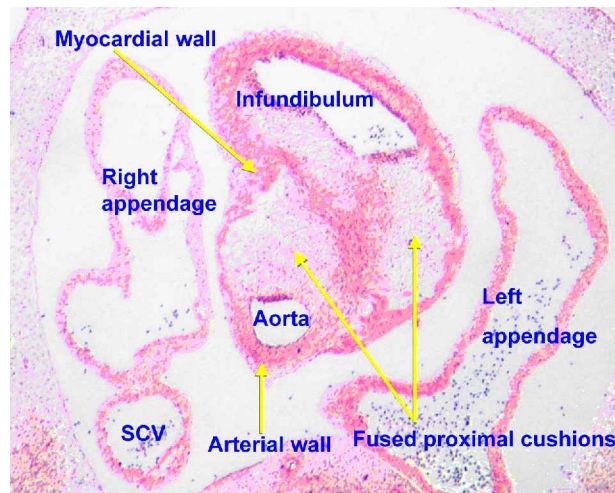
The described morphological findings can be difficult to appreciate in the intact, beating heart, particularly with superimposed changes from long-standing hemodynamic trauma. It can also be difficult to conceptualize the location of the tunnels relative to the subaortic and subpulmonary outflow tracts. With increasing experience, however, we are beginning to



**Figure 15.**

This section, in frontal plane, is from human embryo #13 from the Hamilton collection, estimated to be at Carnegie stage 12. It shows the undivided outflow tract, with its myocardial walls, extending from the roof of the developing right ventricle towards the aortic sac. Note the cushions extending throughout the cavity of the undivided tract, and the bend that divides it into proximal and distal portions. This bend will, eventually, become the sinutubular junction. Reproduced by kind permission of Prof Nigel Brown and Dr Sandra Webb, St George's Hospital Medical School, London.

appreciate the potential embryological origin of the abnormal channels, albeit that none have yet been identified during the stages of their formation. Initially during its development, the solitary outflow tract of the heart has discrete proximal and distal portions (Fig. 15). The junction between these parts will become the sinutubular junction. At first, the entire wall of the outflow tract is composed of myocardium<sup>97,98</sup>. As it is divided by the distal cushions to form the intrapericardial portion of the aorta and the pulmonary trunk, the walls of the distal outflow tract, along with the walls formed from the cushions themselves, transdifferentiate to become arterial structures.<sup>97</sup> The initial myocardial wall persists for a longer period around the proximal outflow tract. Within this part, proximal to the developing sinutubular junction, the distal ends of the cushions that are dividing the outflow tract (Fig. 16), along with the intercalated cushions, transform themselves into the developing arterial sinuses and valvar leaflets. Thus, the cushions that initially fused to septate the proximal outflow tract give rise to the facing sinuses and valvar leaflets of both the aorta and

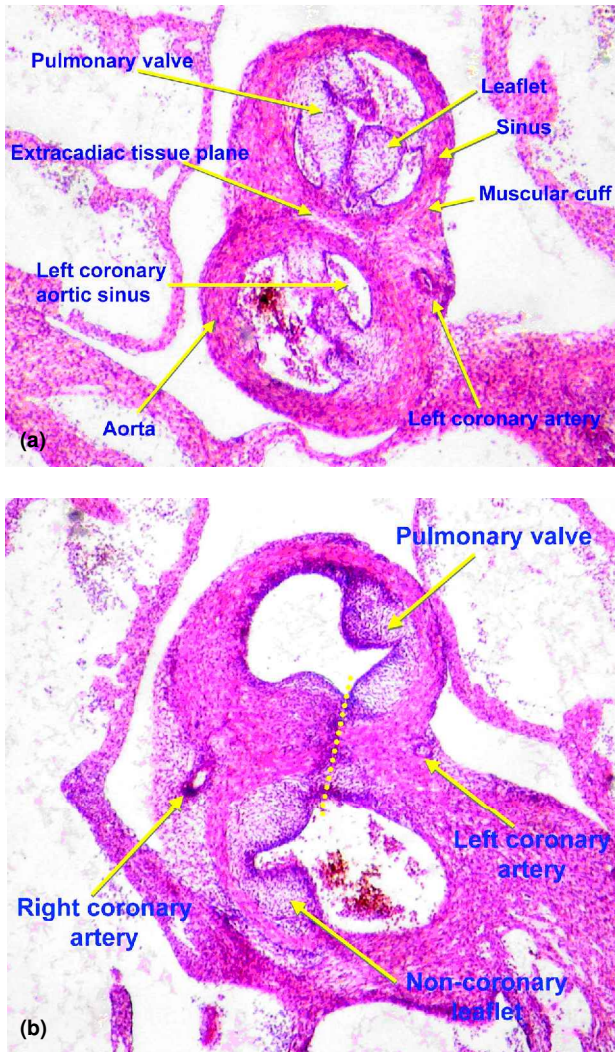


**Figure 16.**

This section, taken in short axis across the proximal outflow tract, is from human embryo #8 in the Hamilton collection, estimated to be at Carnegie stage 16. It shows the developing aortic and pulmonary valvar roots, at this stage encased within a continuous myocardial cuff. Note the muscular tissue growing into the "septal" endocardial cushions. Reproduced by kind permission of Prof Nigel Brown and Dr Sandra Webb, St George's Hospital Medical School, London.

the pulmonary trunk (Fig. 17). The proximal part of the fused cushions, however, hangs down proximal to the forming sinuses as a shelf within the right ventricle (Fig. 18). This most proximal part of the fused cushions then muscularises, initially producing an embryonic outlet septum within the right ventricle (Fig. 18b). With subsequent growth, however, the muscular structure widens to become the free-standing infundibulum of the right ventricle (Fig. 19). At the same time, the cushions lose their septal location, as a tissue plane is formed between the developing roots of the aortic and pulmonary valves (Fig. 17a). In normal development, as the cushions have become converted into the sinuses of the aortic and pulmonary valves, so the myocardial cuff surrounding them has regressed. The disappearance of the muscular cuff causes the tissue plane developing between the infundibulum and the aortic sinuses to lie in communication with the extracardiac space. It is within this tissue plane that abnormal development will produce the aorto-ventricular tunnels, which persist as anomalous channels joining the distal and proximal parts of the initial solitary outflow tract. The precise mechanisms of formation have yet to be clarified, but are probably related to the mechanism of formation of the triangles of fibrous tissue that separate the aortic sinuses beneath the leaflets of the aortic valve, along with abnormal formation of one of the leaflets of the aortic valve. It is also noteworthy that the coronary arteries were initially

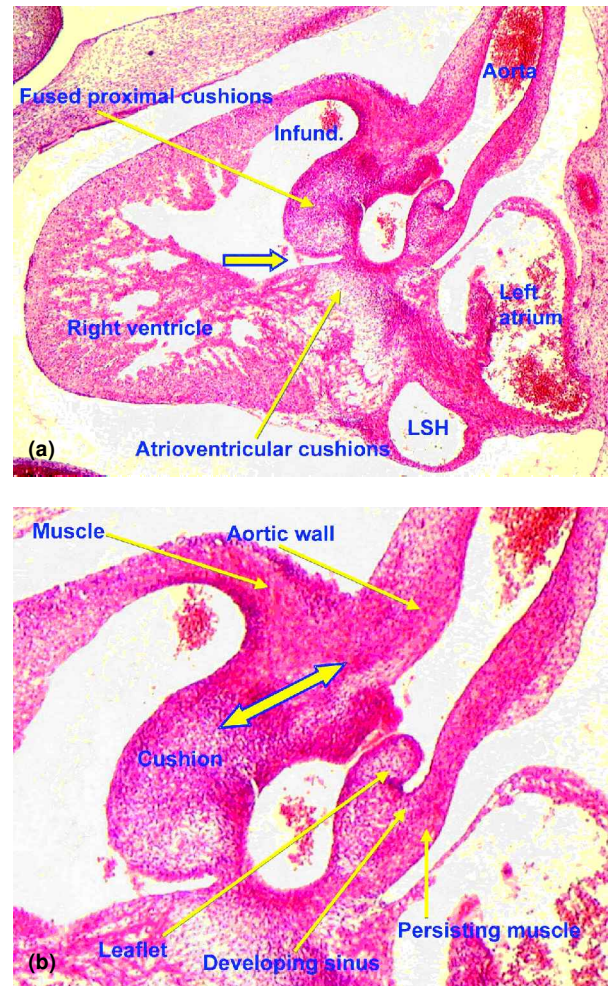




**Figure 17.**

Human embryo #21 from the Hamilton collection is estimated to be at Carnegie stage 20. These sections from the embryo show the developing aortic and pulmonary valvar sinuses and their supporting ventricular roots. Figure (a) is cut obliquely across the sinotubular junction, showing the aortic wall, a small part of the left coronary aortic sinus, and the three leaflets of the developing pulmonary valve, the latter all still encased within a muscular cuff. Note the tissue plane developing between the walls of the aorta and pulmonary trunk. Figure (b) is taken more proximally, and shows how the cushions have fused and muscularised. The dotted line shows their plane of fusion. Again note the muscular cuff which still encases the developing aortic sinuses. Regression of the muscular cuff will place the tissue plane developing between the arterial roots in communication with extracardiac space. The tunnels form abnormally within this tissue plane. Reproduced by kind permission of Prof Nigel Brown and Dr Sandra Webb, St George's Hospital Medical School, London.

encased within the myocardial cuff that surrounded the developing sinuses<sup>97</sup>. The arteries pierce this cuff as they grow into the aortic sinuses.<sup>99,100</sup> It is easy to envisage, therefore, that abnormal development of this junctional region permits channels to form so as to



**Figure 18.**

A sagittal section, replicating the parasternal long axis echocardiographic plane, has been selected from human embryo #17 from the Hamilton collection, estimated to be at Carnegie stage 18. The proximal part of the fused outflow cushions hang as a comma-shaped shelf within the cavity of the right ventricle (a). The arrow points to the closing embryonic interventricular communication. As shown in the enlargement (b), at this stage the muscularising cushions form a right ventricular outlet septum. A tissue plane will eventually form within the shelf to separate the subpulmonary infundibulum from the aortic valvar sinuses as indicated by the arrow, (see Fig. 6). Abnormal formation of this plane permits the development of the aorto-ventricular tunnels. Reproduced by kind permission of Prof Nigel Brown and Dr Sandra Webb, St George's Hospital Medical School, London.

connect the aorta with the outflow tract of the left ventricle, by-passing the attachment of the valvar leaflet to produce an aorto-left ventricular tunnel, or with the infundibulum, giving rise to the aorto-right ventricular tunnel. We presume that the abnormal development involves failure of the outflow cushions properly to form the arterial sinuses, the valvar leaflets, and the fibrous interleaflet triangles, coupled

with abnormal separation of the distal outflow tract into the aorta and pulmonary trunk. At the same time, the muscularising proximal cushions become converted into the proximal myocardial wall of the tunnel. The associations of the tunnels with abnormalities of the aortic sinuses, the proximal coronary arteries, and the leaflets themselves, therefore, are entirely predictable. The end-result is one of the few cardiac malformations in which congenital lesions

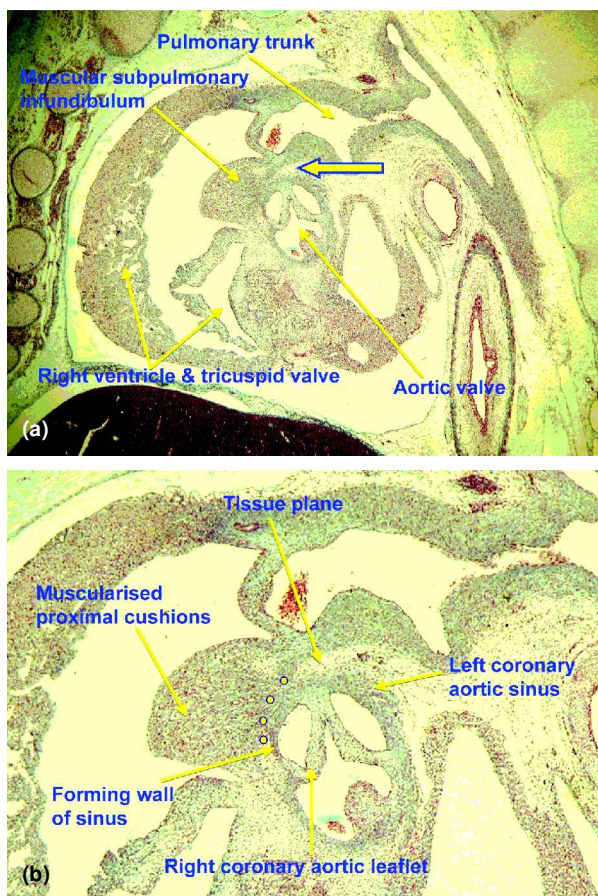
of both aortic and pulmonary valvar leaflets may coexist.<sup>1,26</sup>

### Clinical presentation

The most consistent echocardiographic feature on antenatal examination between 18 and 33 weeks gestation is dilation and hypertrophy of the left ventricle, with severe and progressively reduced shortening fraction. Apparent aortic regurgitation, which is extremely uncommon during fetal life, and enlargement of the aortic root, further support a diagnosis of aorto-ventricular tunnel, while flow of blood around the hinge of the valve has been imaged with color flow Doppler echocardiography.<sup>43</sup> Thickening, or severe dysplasia, of the aortic valvar leaflets was found in three of seven fetal cases, suggesting that this group may represent the more severe end of the pathological spectrum. Furthermore, an incidence of 0.46% among fetal cardiac malformations<sup>42</sup> is nearly five times greater than has been previously recognized after birth.<sup>25</sup>

At birth, or at the initial examination, there is invariably a loud "to-and-fro" murmur. This is usually accompanied by systolic and diastolic thrills, and is heard over the entire precordium. Bounding peripheral pulses are also a consistent finding. Enlargement of the heart, and uniform dilation of the ascending aorta, are usually obvious on chest X-ray, although the dilated aorta is not infrequently mistaken for the thymus gland. The electrocardiogram occasionally is normal,<sup>27</sup> but typically shows left or biventricular hypertrophy, with a "strain pattern" of inverted T waves seen in the precordial leads. Although clinical signs in older patients closely mimic those of valvar aortic stenosis and incompetence, with a widened pulse pressure, and systolic and diastolic murmurs, the aortic component of the second heart sound is conserved, as is a dicrotic notch on the arterial pressure trace.<sup>6</sup> Cardiac enlargement is seen on the chest X-ray, which can also reveal marked enlargement of the entire ascending aorta. With the passage of time, the dilation can become extreme, and may appear disproportionate to other signs and symptoms of cardiac disease. In some patients, the tunnel itself can be seen as a leftward prominence of the aortic root in the area of the pulmonary trunk.<sup>6</sup>

The onset, and severity, of symptoms is highly variable, and probably reflects complex interactions and incompletely understood contributions from the lesion itself, the compromised coronary circulation, and any associated malformations. While occasional patients remain active and asymptomatic into adulthood,<sup>7,27,35,45</sup> there are also reports of fetal death,<sup>43</sup> as well as rapidly fatal congestive heart failure<sup>23,31</sup> and sudden death<sup>40</sup> in previously compensated children



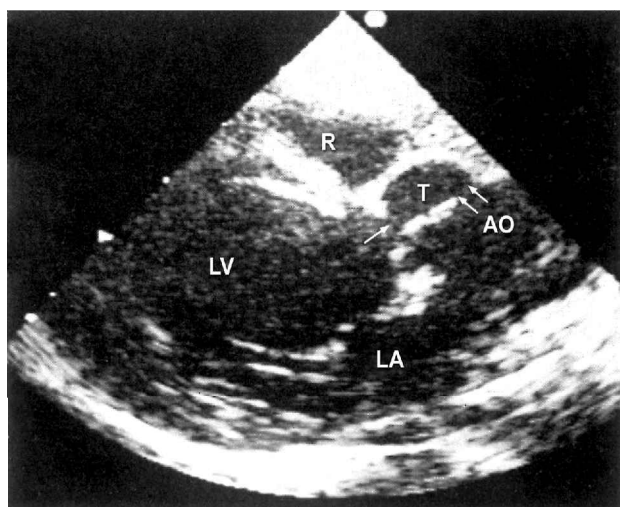
**Figure 19.**

*This section, again in sagittal plane replicating the parasternal long axis echocardiographic cut, is from another human embryo from the Hamilton collection, estimated to be at Carnegie stage 22, just after the completion of cardiac septation. The comma-shaped muscular shelf that, at stage 20, formed an intraventricular muscular outlet septum, has now been transformed into the muscular subpulmonary infundibulum (a). Note the tissue plane already formed between the pulmonary trunk and the aorta (arrow). The enlargement (b) shows that the tissue plane has yet to extend to separate the forming wall of the right coronary aortic sinus from the infundibulum, itself formed by muscularisation of the proximal cushions which divided the outflow tract. It is precisely within the site of formation of this tissue plane (dotted line) that we find the abnormal aorto-ventricular tunnels. Reproduced by kind permission of Prof Nigel Brown and Dr Sandra Webb, St George's Hospital Medical School, London.*

and adults. These latter groups may have a higher incidence of coronary arterial compromise, with or without obstruction of the right ventricular outflow tract. The majority of patients suffer congestive heart failure within the first year of life, and many show this feature during the neonatal period. This is true whether the tunnel communicates with the left or with the right ventricle, although pulmonary stenosis in association with aorto-right ventricular tunnel may delay the onset of symptoms.<sup>91</sup> Attempts have been made to correlate the clinical course with morphology of the tunnel itself,<sup>41,101</sup> but information in the literature is probably inadequate presently to substantiate meaningful inferences. The association with severe dysplasia, stenosis, or atresia of the aortic valve, nonetheless, constitutes a particularly lethal combination of malformations. Of eleven such cases,<sup>16,32,43,52,53,60,72,76,77,86</sup> four died before birth or on the first day of life, with the remaining patients all developing congestive heart failure or low cardiac output early in the neonatal period.

### Investigation

Echocardiography, with cross-sectional and color-Doppler imaging, constitutes the diagnostic investigation of choice.<sup>14,15,28,34</sup> The parasternal long-axis view shows the tunnel beside the aorta, from which it can be followed to its aortic and ventricular openings. As explained, when opening to the left ventricle, these are above and below the right or left coronary

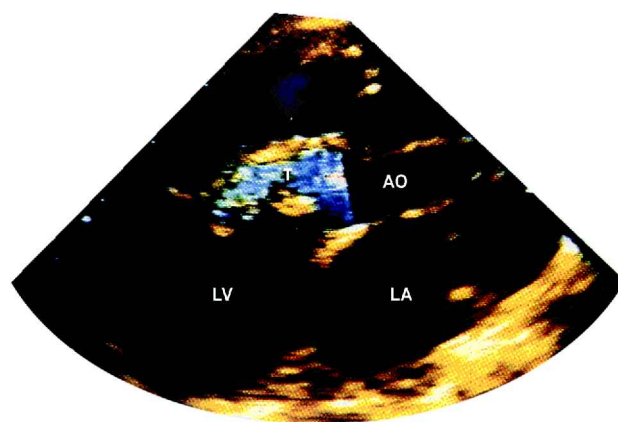


**Figure 20.**

The cross-sectional echocardiogram in the parasternal long axis view shows a tunnel (T) between the aorta (AO) and the left ventricle (LV). Small arrows indicate the aortic and ventricular orifices of the tunnel. Reproduced from Sreeram N, Franks R, Walsh K. Aortico-left ventricular tunnel: long-term outcome after surgical repair. *J Am Coll Cardiol* 1991; 17: 950–955, with permission from Elsevier Science. Abbreviations: R: right ventricle; LA: left atrium.

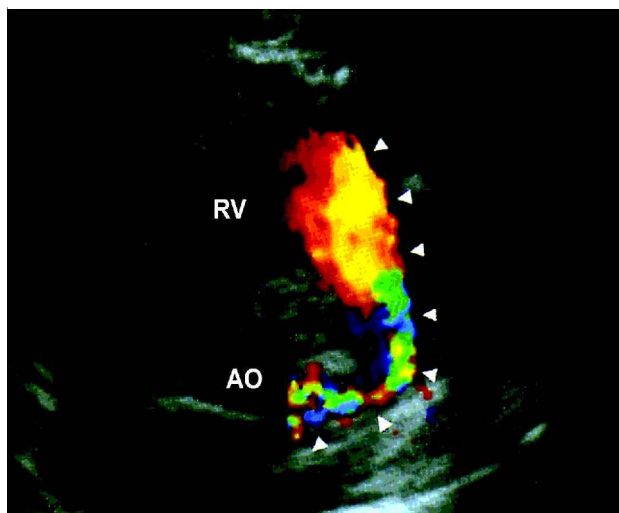
aortic sinuses, respectively (Fig. 20). Color-flow imaging demonstrates blood passing through the abnormal channel from the left ventricle to the aorta during systole, and in the opposite direction in diastole (Fig. 21). The right ventricular outflow tract, and the pulmonary valve, are also seen. This permits quantification of any obstruction due to displacement of the subpulmonary infundibulum, and identification of the much rarer tunnel extending from the aorta to open in the right ventricle.<sup>36,86,90</sup> Parasternal short-axis views at the levels of the aortic valve and ascending aorta should show intact, albeit often enlarged, sinuses of Valsalva. These views also reveal any thickening or dysplasia of the valvar leaflets, the coronary arterial origins, and typically show uniform enlargement of the ascending aorta, which may be twice its normal diameter.<sup>21</sup> The short axis views also demonstrate tunnels opening to the right ventricle (Fig. 22).<sup>90</sup> In the apical four-chamber view, left ventricular dilation and hypertrophy, with variable impairment of the shortening fraction, and otherwise conserved cardiac architecture, are characteristic.

Of all these features, extensive and uniform dilation of the ascending aorta may be the best non-invasive clue to the diagnosis of aorto-ventricular tunnel, for this is hardly ever present early in life with other cardiac malformations. Only extremely rarely is enlargement of the aorta not present, specifically when there is critical obstruction both to the aortic valve and within the tunnel.<sup>72</sup> The most common diagnostic errors from echocardiography have been to confuse the ventricular end of the tunnel with a ventricular septal defect,<sup>1,26</sup> or to mistake displacement of the



**Figure 21.**

Doppler colour-flow imaging shows diastolic flow through a tunnel from the aorta to the left ventricle. Abbreviations: LA: left atrium, LV: left ventricle, T: tunnel, AO: aorta. Reproduced from Sousa-Uva M, Touchot A, Fermont L, Piot D, Delezoides AL, Serraf A, Lacour-Gayet F, Roussin R, Bruniaux J, Planché C. Aortico-left ventricular tunnel in fetuses and infants. *Ann Thorac Surg* 1996; 61: 1805–1810, with permission from Elsevier Science.

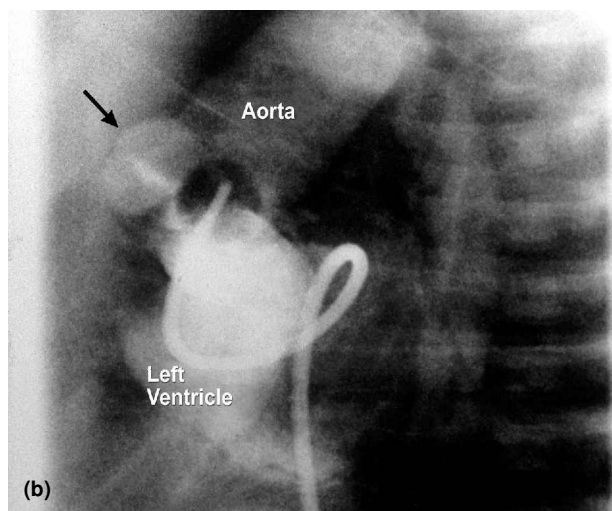
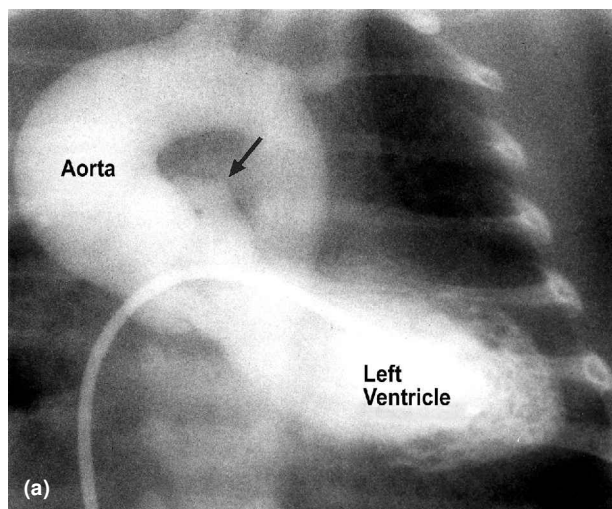


**Figure 22.**

A parasternal short axis echocardiogram with Doppler colour flow mapping shows a tunnel (arrow beads) extending from the aorta to open into the right ventricle in a ten-day-old neonate. The patient also had valvar pulmonary stenosis. The external appearances of this heart are shown in Figure 8. Abbreviations: AO: aorta; RV: right ventricle. Reproduced from Hrudá J, Sobotka-Plojhar MA, van Rossum AC. Aortico-right ventricular tunnel with pulmonary stenosis in a neonate. *Heart* 2001; 86: 316, with permission from BMJ Publishing Group.

subpulmonary infundibulum for Fallot's tetralogy.<sup>42</sup> Flow of blood through the tunnel has also been misinterpreted as valvar aortic regurgitation,<sup>42</sup> or a ruptured aneurysm of the sinus of Valsalva.<sup>43</sup>

Although previously considered essential in the investigation of suspected aorto-ventricular tunnel, the present role of cardiac catheterization is to clarify the coronary arterial anatomy when the proximal vessels cannot reliably be imaged by echocardiography, and possibly to elucidate some associated malformations. Hemodynamic studies routinely confirm normal pressures in the right heart, even in the presence of massive left ventricular enlargement,<sup>6,29,36</sup> unless the tunnel has compressed the right ventricular outflow tract.<sup>1,31,36</sup> Typical findings in the left heart are a widened aortic pulse pressure, with a normal or minimally elevated left ventricular end diastolic pressure.<sup>29</sup> Although gradients have been documented across the left ventricular outflow tract,<sup>1,6</sup> flow through the tunnel itself may conceal significant aortic valvar obstruction.<sup>30,72</sup> The shape and extracardiac course of the tunnel can be demonstrated by angiography (Fig. 23), as can obstruction of the subpulmonary infundibulum (Fig. 24), but this information adds little to that obtained from high-quality sector scanning. Magnetic resonance imaging<sup>34,90</sup> also shows clearly the structure of the tunnel and its anatomical



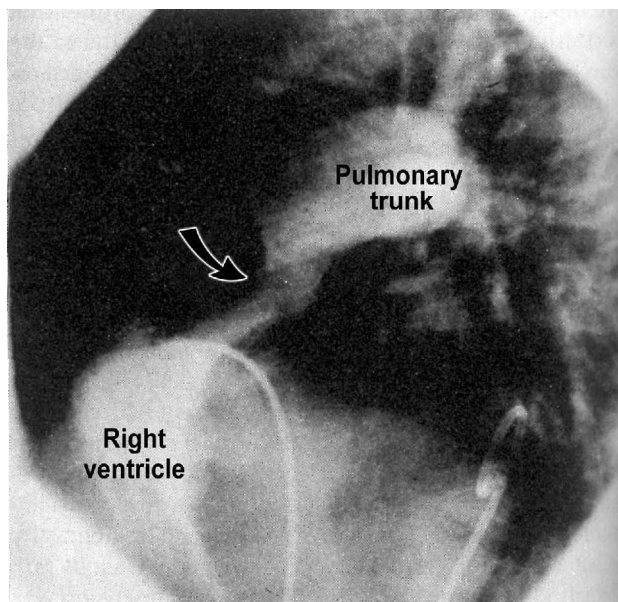
**Figure 23.**

A left ventriculogram, profiled in the antero-posterior view (a), and an ascending aortogram, profiled in lateral projection (b), show a tunnel (arrow) extending from the aorta to the left ventricle in a neonate.

relationships, but remains of limited availability in most clinical situations. Both resonance imaging and interventional catheterization, however, could, potentially have wider application for management of these patients, the former to characterize abnormal patterns of flow in the aortic root, and the latter to relieve valvar obstruction.<sup>50,79,90</sup>

### Differential diagnosis and associated malformations

A number of other cardiac malformations must be excluded from the differential diagnosis of patients with congestive heart failure and signs of aortic regurgitation (Table 1). In the neonate or young infant, a ruptured fistula of the sinus of Valsalva, ventricular



**Figure 24.**

*In this patient, a right ventricular angiogram demonstrates compression of the subpulmonary infundibulum (arrow) by a tunnel extending from the aorta to the left ventricle in a five-year-old patient. Reproduced from Knott-Craig CJ, van der Merwe PL, Kalis NN, Hunter J. Repair of aortico-left ventricular tunnel associated with subpulmonary obstruction. *Ann Thorac Surg* 1992; 54: 557-559, with permission from Elsevier Science.*

**Table 1.** Differential diagnosis of aorto-ventriculartunnel.

Sinus of valsalva fistula
Common arterial trunk with valvar regurgitation
Aorto-pulmonary window
Ventricular septal defect with aortic regurgitation
Persistent patency of the arterial duct
Coronary-cameral fistula
Tetralogy of Fallot with absent pulmonary valve
Valvar aortic stenosis and regurgitation
Cerebral arterio-venous malformation

septal defect with aortic regurgitation, and valvar aortic incompetence are extremely uncommon. Furthermore, they do not generally have accompanying aortic dilation with left ventricular dysfunction. The murmurs of the persistently patent arterial duct, aorto-pulmonary window, and coronary-cameral fistulas are more continuous than “to-and-fro” in nature, while that of a cerebral arterio-venous malformation tends to localize over the head. Echocardiography should readily distinguish cases of common arterial trunk and Fallot’s tetralogy. In older patients, the history of loud systolic and diastolic murmurs heard soon after birth favors a diagnosis of aorto-ventricular tunnel, as does disproportionate enlargement of the heart and ascending aorta.

**Table 2.** Cardiac anomalies associated with aorto-ventricular tunnel in 105 reported cases.

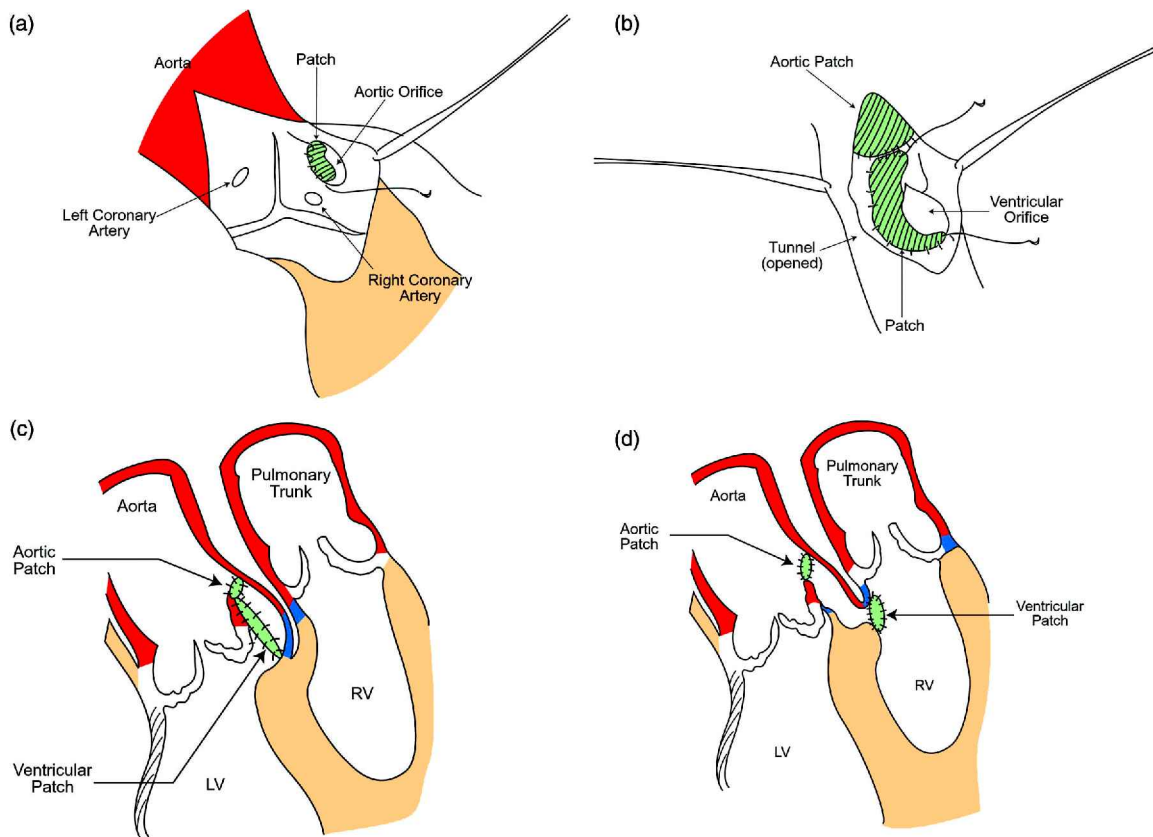
Cardiac lesion	Number of cases	References
Aortic valve		
2-leaflet	6	1, 3, 21, 26, 31, 40
Obstructed 3-leaflet	5	6, 7, 41, 42, 59
Dysplastic/atretic	10	1, 16, 30, 32, 43, 52, 53, 60, 72, 76
Perforation of leaflet	2	8, 9
Unspecified regurgitation	2	21, 45
Valvar pulmonary stenosis	7	1, 26, 50, 78, 83, 87, 90, 91
Dysplastic tricuspid valve	1	83
Absent origin of coronary artery		
Right	6	6, 21, 31, 41, 49, 87
Left	1	88
Aneurysm of sinus of Valsalva	3	17, 30, 85
Atrial septal defect/patent oval foramen	8	31, 37, 38, 41, 72, 83, 84, 85
Ventricular septal defect	3	15, 21, 38
Persistent patency of the arterial duct	19	15, 16, 20, 21, 26, 30, 31, 38, 41, 52, 60, 72, 79, 83, 84, 89
Absent left atrial appendage	1	40

Associated anomalies, apart from those involving the aortic sinuses, the aortic and pulmonary valvar leaflets, and the coronary arteries, have been found infrequently among this group of patients (Table 2). No associations have been observed with any genetic syndromes or extracardiac defects, but reports of aorto-ventricular tunnel are rare among patients of African, Oriental, or Asian descent. An approximate ratio of two males to one female has remained constant among reported cases

## Treatment

While it has been proposed that congenital “aortic regurgitation” is better tolerated than that acquired later in life,<sup>35</sup> there is no evidence to support medical treatment for patients having an aorto-ventricular tunnel. Without surgical intervention, most die early in life from congestive heart failure.<sup>9,10,41</sup> Only patients undergoing surgery before six months of age have later had documentation of normal left ventricular size and function.<sup>43</sup> Moreover, lack of support for the right or left coronary aortic leaflet invariably results in progressive aortic regurgitation,<sup>102</sup> often necessitating repair or replacement of the valve as a primary<sup>45</sup> or secondary<sup>7,8,57,58,73</sup> procedure. Surgery, therefore, should be undertaken without delay, even in asymptomatic patients.<sup>25,103</sup>

The goals of surgery are complete closure of the abnormal communication, restoration of aortic valvar function, preservation of the coronary arterial



**Figure 25.**

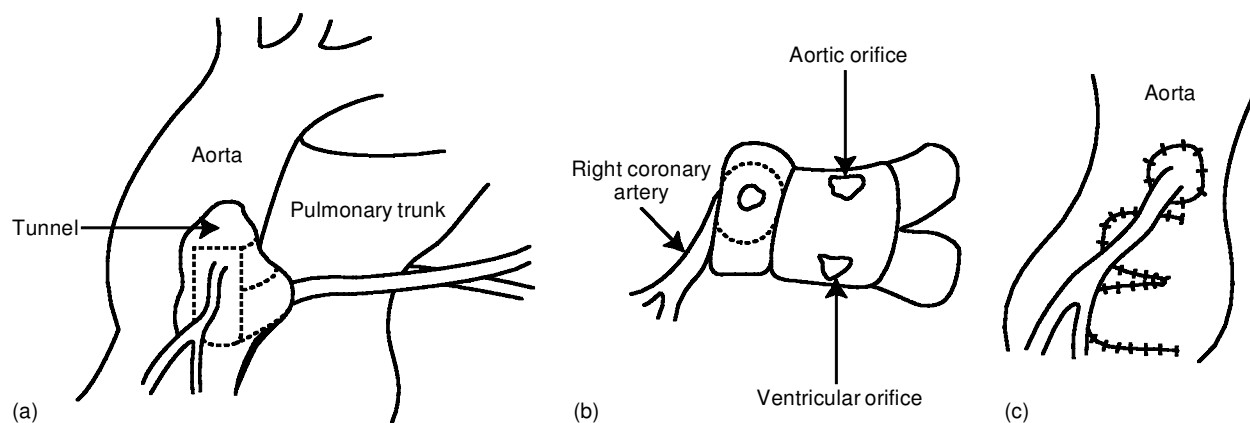
*These diagrams illustrate the most-commonly employed technique for repair of an uncomplicated tunnel running from the aorta to the left ventricle (a,b,c) or the right ventricle (a,d). The aortic orifice (a) is closed through an aortotomy by suturing a patch to the sinutubular ridge and the aortic wall, having identified the orifices of both coronary arteries. The tunnel itself is opened vertically (b). The ventricular orifice is closed using a second patch, which is sutured to the ventricular myocardium, and, in the case of tunnels ending within the left ventricle, also to the fibrous wall of the unsupported aortic sinus, and the bottom of the first patch. The walls of the tunnel are then approximated over the patches, and the aortotomy is closed. Figure (c) shows the completed repair, with the aortic leaflet now supported by the patches. Figure (d) shows a completed repair for a tunnel terminating in the right ventricle. Drawn after Ho et al.<sup>92</sup>*

circulation, and relief of any obstruction within the right or left ventricular outflow tracts. Transcatheter closure of a tunnel to the left ventricle with an Amplatzer duct occluder has been reported in a single patient.<sup>52</sup> Attempted coil closure of a tunnel to the right ventricle, however, was not successful.<sup>90</sup> Considering the benefit of providing support for the aortic valvar leaflets, as well as the spectrum of associated coronary arterial anomalies, it seems likely that repair of the aorto-ventricular tunnels should remain largely, if not exclusively, within the surgical domain.

Surgical repair of an uncomplicated tunnel to the left ventricle is performed on cardiopulmonary bypass, usually using a single right atrial cannula and moderate hypothermia. The tunnel is compressed externally immediately after commencement of perfusion, and during administration of cardioplegia, thus preventing ventricular distension and facilitating myocardial protection, respectively. Origin of a coronary artery deep within the tunnel could be an indication for retrograde delivery of cardioplegia through the coronary sinus.

While simple suture of the aortic end of the tunnel, essentially approximating the sinutubular ridge to the aortic wall, has occasionally given good results,<sup>13,17</sup> it is generally accepted that a patch should be used to avoid further distortion of the aortic valvar leaflet.<sup>10,20,21</sup> This is done through a transverse or oblique aortotomy, using a continuous monofilament suture to attach a Gore-Tex or pericardial patch to the sinutubular ridge and aortic wall (Fig. 25a).

When the orifice of a coronary artery lies within the proximal part of the tunnel, the patch on the aortic wall is deviated distally to avoid its exclusion. In all cases, care is taken to avoid narrowing a coronary arterial orifice that lies close to the aortic orifice of the tunnel, or a branch that follows an intraluminal course in its wall. The ventricular end of a tunnel is closed with a second patch placed through a vertical incision into the tunnel itself (Fig. 25b). This both provides support the right aortic sinus, and prevents ongoing displacement of the right ventricular outflow tract by high-pressure and turbulent flow in a blind-ending pouch.<sup>36</sup> The bottom of this patch is



**Figure 26.**

*These diagrams illustrate an alternative technique for repair of a tunnel opening into the left ventricle. The wall of the tunnel is opened vertically, in this instance leaving a generous margin of tissue around the right coronary artery, which arises distally from the tunnel. The orifice of the right coronary artery is excised with a surrounding button of tunnel wall (a), and the opposite flap of tunnel tissue is incised to the wall of the aorta. Both the aortic and ventricular openings can be seen from the tunnel (b). The wall of the tunnel is now sutured around each orifice as a patch, closing the aortic and ventricular ends as well as supporting the right coronary aortic leaflet and obliterating the lumen of the tunnel. The right coronary artery is reimplanted into the ascending aorta above the closed orifice of the tunnel (c). The dashed lines indicate surgical incisions. Drawn after Grünenfelder et al.<sup>47</sup>*

sutured to the ventricular myocardium, while the top, of necessity, must be joined to the first patch and the wall of the aortic sinus (Fig. 25c,d). In a single case, an elegant plastic repair (Fig. 26a–c) used the tunnel itself to achieve complete obliteration of the abnormal channel and stabilization of the aortic valve, avoiding the need to implant artificial material.<sup>47</sup> The ventricular end of a tunnel opening to the right ventricle is also usually closed either with a patch, or by direct suture<sup>78,80</sup> through a right ventriculotomy. On occasion, both the aortic and ventricular ends have been patched through the tunnel.<sup>79</sup> In theory, however, tunnels to the right ventricle should not jeopardize support for the aortic leaflet, and the ventricular end of the tunnel is at the same pressure as the subpulmonary infundibulum. It is questionable, therefore, whether closure of the ventricular orifice of a tunnel ending in the right ventricle is either useful or necessary.

Should the right coronary artery originate distally within a tunnel arising above the right coronary aortic sinus, it is resected with a generous button of the surrounding wall of the tunnel and anastomosed to the ascending aorta.<sup>20,21,47,49</sup> While similar treatment of the anterior interventricular<sup>78</sup> or circumflex<sup>85</sup> branches of the left coronary artery would seem logical, this has not, as yet, been reported. A left coronary artery, or one of its major branches, taking origin distally from a tunnel that originates above the left sinus of Valsalva presents a more difficult problem. Owing to its position in the transverse sinus, and its distance from the aorta, transfer of the coronary artery to the aorta may be technically more complicated or impossible. This situation has been encountered, thus

far, mainly in tunnels terminating in the right ventricle, where closure of the ventricular, rather than the aortic, end of the tunnel has successfully conserved coronary arterial perfusion.<sup>91</sup> Presence of such an arrangement with a tunnel entering the left ventricle might constitute an indication for leaving open the ventricular end of the tunnel. Intraoperative differentiation of absence of the left coronary arterial orifice<sup>88</sup> from origin deep within the tunnel<sup>84,91</sup> may be impossible. This fact emphasizes the need to obtain accurate imaging of the coronary arterial origins preoperatively. A stenotic coronary arterial orifice, if recognized during life, could be an indication for grafting with the internal mammary artery.

Valvar pulmonary stenosis in association with tunnels entering either the left<sup>26,50</sup> or the right ventricle<sup>86,90,91</sup> has successfully been managed by open valvotomy<sup>26,86,91</sup> or preoperative percutaneous balloon valvoplasty,<sup>50,57</sup> although balloon dilation did not achieve satisfactory relief of the obstruction in one case.<sup>90</sup> Aortic valvar lesions are treated on their own merits. Commissurotomy for stenotic valves with two and three leaflets,<sup>32,53,72</sup> homograft replacement of the aortic root,<sup>76</sup> and, in a single case of aortic atresia, neonatal aortoventriculoplasty<sup>52</sup> have all been employed in small infants, as well as valvar repair or replacement in older children and adults.

## Results

Survival following surgical repair has improved steadily. Prior to 1983, there was a collective mortality of 20%.<sup>10</sup> In recent series, mortality has approached zero.<sup>21</sup> While initial reports indicated a very high

incidence of severe aortic valvar regurgitation occurring eight to twelve years after operation,<sup>7,8</sup> most of these patients had been repaired by direct suture of the aortic end of the tunnel, and came to surgery beyond five years of age. A shorter period of follow-up now suggests that patients in whom the aortic sinus is supported, and the tunnel closed in early infancy, should have little if any leakage through their aortic valve, as well as a good possibility of normal left ventricular function.<sup>21,43</sup> The subset of patients with associated dysplasia, stenosis, or atresia of the aortic valve, however, remains a significant challenge. Although aggressive relief of obstruction within the left ventricular outflow tract at the time of repair has achieved some success, overall surgical mortality remains close to 50% in this cohort, and most survivors require multiple replacements of the aortic valve.<sup>52,76</sup> The extremely rare origin of a left coronary artery within a tunnel arising above the left aortic sinus may also constitute a surgical risk factor, as this combination is not readily apparent at the time of operation, and closure of the aortic orifice of such a tunnel extending to the right ventricle has proved fatal.<sup>84</sup>

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