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Echocardiographic assessment of the aortic valve and left ventricular outflow tract

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THE PREVIOUS REVIEWS IN THIS SECTION OF OUR Supplement[1,2] have summarized the anatomic components of the ventriculo-arterial junctions, and then assessed the echocardiographic approach to the ventriculo-arterial junction or junctions as seen in the morphologically right ventricle. In this complementary review, we discuss the echocardiographic assessment of the comparable components found in the morphologically left ventricle, specifically the outflow tract and the arterial root. We will address the echocardiographic anatomy of the aortic valvar complex, and we will review the causes of congenital arterial valvar stenosis, using the aortic valve as our example. We will also review the various lesions that, in the outflow of the morphologically left ventricle, can produce subvalvar and supravalvar stenosis. We will then consider the salient features of the left ventricular outflow tract in patients with discordant ventriculo-arterial connections, and double outlet ventricles. To conclude the review, we will briefly address some rarer anomalies that involve the left ventricular outflow tract, showing how the transesophageal echocardiogram is used to assist the surgeon preparing for repair. The essence of the approach will be to consider the malformations as seen at valvar, subvalvar, or supravalvar levels,[1] but we should not lose sight of the fact that aortic coarctation or interruption, hypoplasia of the left heart, and malformations of the mitral valve are all part of the spectrum of lesions associated with obstruction to the left ventricular outflow tract. These additional malformations, however, are beyond the scope of this review.

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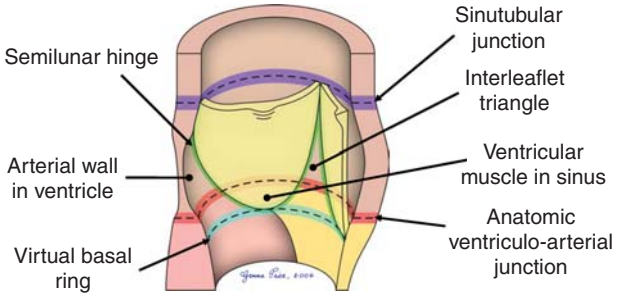

Figure 1.
The cartoon shows the essential anatomic features of the arterial root. Note that none of the illustrated rings represents the surgical "annulus", which is the remnants of the semilunar hingepoints seen when the surgeon has removed the diseased leaflets from the arterial root.

Echocardiographic anatomy

As emphasised in the anatomic review,[1] the left ventricular outflow tract consists of the arterial valve, usually the aortic valve but the pulmonary valve when the ventriculo-arterial connections are discordant, and its supporting ventricular structures. These supporting structures can be divided into an antero-superior component, made up of the muscular ventricular septum and the parietal left ventricular wall, and a postero-inferior component, which is almost always fibrous, being made up of the area of fibrous continuity between the leaflets of the arterial valve and the mitral valve, along with the membranous septum.

Although it still remains conventional to describe an "annulus" within the arterial valve, the echocardiographer needs to be aware that, in reality, the arterial root possesses at least three discrete rings, mainly the anatomic ventriculo-arterial junction, the sinutubular junction, and the virtual basal ring, but that none of these rings supports the leaflets of the arterial valve in annular fashion (Fig. 1). Instead, it is the

semilunar attachments of the three arterial valvar leaflets, interdigitating with the arterial sinuses, which produce three fibrous inter-leaflet triangles that extend to the level of the sinutubular junction. The semilunar hinges of the normal valve permit the leaflets to open and close along three discrete zones of apposition, these extending from the centre of the valvar orifice to the sinutubular junction. It is the peripheral attachments of these zones of apposition that are usually described as the three valvar commissures. In most instances, the leaflets of the valve are of different sizes.[3] When the left ventricle supports the aorta, then usually the noncoronary leaflet is the largest. The hinge-point of all three leaflets within the root is elliptical and, corresponding to the midpoint of each leaflet, the aortic wall bulges outwards to form the sinuses of Valsalva. It is the remains of the semilunar attachments, seen when the surgeon has removed the leaflets, that are usually described as the surgical "annulus", but it is conventional for the echocardiographer to take the lower limits of the sinuses of Valsalva as being the "annulus" when measuring the diameter of the outflow tract. This ring, however, is a virtual locus (Fig. 1), and the dimensions of the root at this level are significantly less than those measured at mid-sinusal level, albeit that the root narrows again at the sinutubular junction, which is another obviously annular structure, although rarely described in this fashion.

Aortic valvar stenosis

Aortic stenosis can be caused by deformities of the valvar leaflets, usually with but sometimes without narrowing of the ventriculo-aortic junction. Stenosis is most frequently the consequence of fusion of the leaflets along the zones of apposition between them. Complete fusion of a putative zone of apposition is termed a raphe. Echocardiographically, it is possible to distinguish such a raphe from a patent zone of apposition during systole, but the two can be confused in diastole, as the echo-bright lines recorded from the raphe are indistinguishable from the images produced by patent zone of apposition. Valvar stenosis can be unicuspid and unicommisural, bicuspid, tricuspid, quadricuspid, or can be produced by the dysplastic leaflets of the so-called undifferentiated aortic valve. In the latter instance, there may not be fusion of the zones of apposition between the abnormal leaflets, the mere bulk of the leaflets being sufficient to obstruct the outflow tract.

The feature of the so-called unicuspid valve is the presence of a single slit-like zone of apposition within a skirt of leaflet tissue, albeit that one, or usually two, aborted zones can be found that present as raphes. The solitary zone of apposition extends only to the centre of the valvar orifice. In systole, therefore, the orifice itself appears circular or oval (Fig. 2), and is eccentrically located.[3] This variant is the commonest cause of critical aortic stenosis as seen in the neonate.

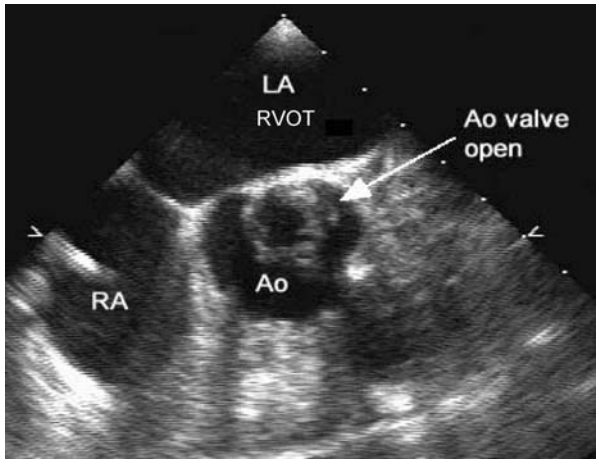

Figure 2.
This short axis cut shows a unicuspid and unicommisural aortic valve. Note the eccentrically located valvar orifice.

The bicuspid aortic valve is the commonest cause of aortic stenosis. Indeed, an aortic valve with two leaflets is said to exist in 2 percent of the general population, albeit that it is not always stenotic. When seen in parasternal short axis cuts, or in the sector from 0 to 30° when interrogated transoesophageally from the mid-oesophagus, the valve can be visualized en-face, showing two leaflets which usually are similar in size. The bicuspid valve with leaflets supported only by two sinuses, a rare variant, has a single line of closure. More commonly, two of the putative leaflets have fused during development, producing an arrangement that may look tricuspid in diastole because of the presence of a raphe. In systole, the leaflets separate along a solitary zone of apposition, and the orifice looks like the mouth of a fish. This is the classical echocardiographic appearance of aortic valve with two leaflets (Fig. 3).

A stenotic aortic valve with three leaflets is more frequently seen in the aged population, where the fused zones of apposition also become calcified. In children, nonetheless, valves can be found with partially fused zones of apposition exacerbated by lumps of fibrous tissue. The edges of the leaflets themselves can be rolled or severely thickened, but typically the leaflets are well-developed. The extent of stenosis depends on the degrees of fusion along the zones of apposition. When seen with a narrowed aorto-ventricular junction, it can be another cause of neonatal aortic stenosis.

An aortic valve with four leaflets is rare. There are various different configurations. The commonest is to find three leaflets of normal size along with a small accessory leaflet. In diastole, closure along the

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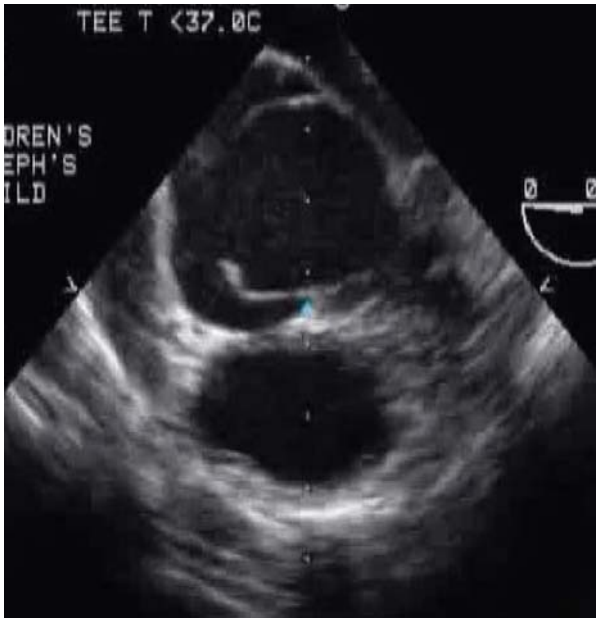

Figure 3.
This short axis cut shows an open aortic valve with only two leaflets.

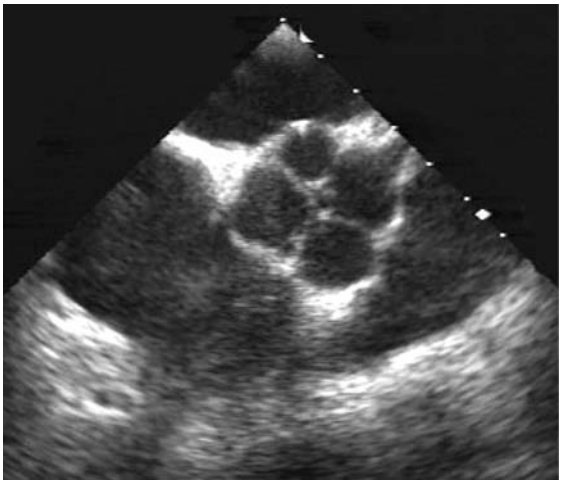

Figure 4.
This arterial valve has four leaflets. It is from a patient with a common arterial trunk, but the same arrangement can rarely be seen in the aortic valve.

zones of apposition produces an X-pattern, as seen more frequently in the common arterial valve with four leaflets (Fig. 4).

The so-called undifferentiated valve is basically a fibrous diaphragm of dysplastic and immobile tissue. This is also a common cause of neonatal critical aortic stenosis.

Critical aortic stenosis

This malformation is a common cause of neonatal congestive heart failure. The leaflets of the valve are usually thickened, with the unicuspid or undifferentiated valves being the commonest anatomic subtype. Left ventricular dysfunction (Fig. 5), and endocardial fibroelastosis, are common, and often the systemic circulation is duct-dependent. It is the size of the mitral valve and the left ventricle that determines whether these patients will be suitable for biventricular repair or univentricular palliation, along with the presence or absence of fibroelastosis.

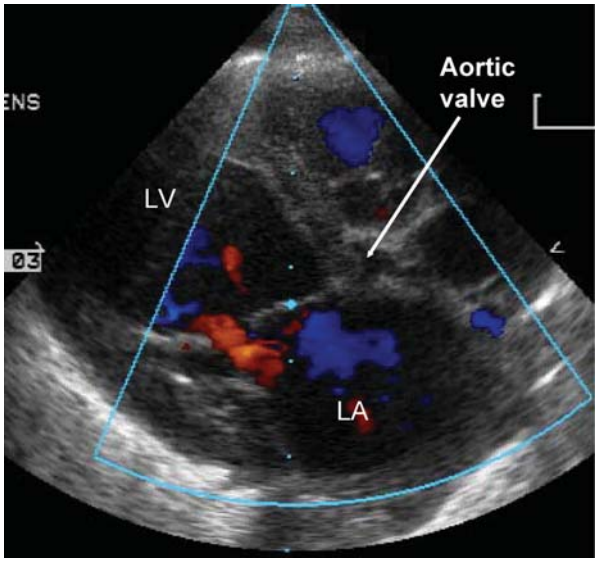

Figure 5.
This patient has neonatal critical aortic stenosis. Note the dilated left ventricle and the thickened aortic valve.

Subvalvar aortic stenosis

Stenosis below the level of the arterial valve can take multiple forms (Fig. 6). These lesions produce subaortic stenosis in the setting of concordant ventriculoarterial connections, but the same malformations result in subpulmonary stenosis when the ventriculoarterial connections are discordant. When producing subaortic stenosis, the commonest lesions are a discrete subaortic fibrous shelf (Fig. 7), a combination of the shelf with muscular stenosis, which can become elongated to form a tunnel (Fig. 8), or hypertrophic obstructive cardiomyopathy (Fig. 9).

Supravalvar aortic stenosis

This may be idiopathic, familial, or may be associated with William's syndrome or congenital rubella syndrome. Three anatomic subtypes have been described. These include the membranous type, which consists of a single simple fibrous diaphragm, the hourglass type, which consists of an internal and external stricture, and the diffuse hypoplastic type (Fig. 10),

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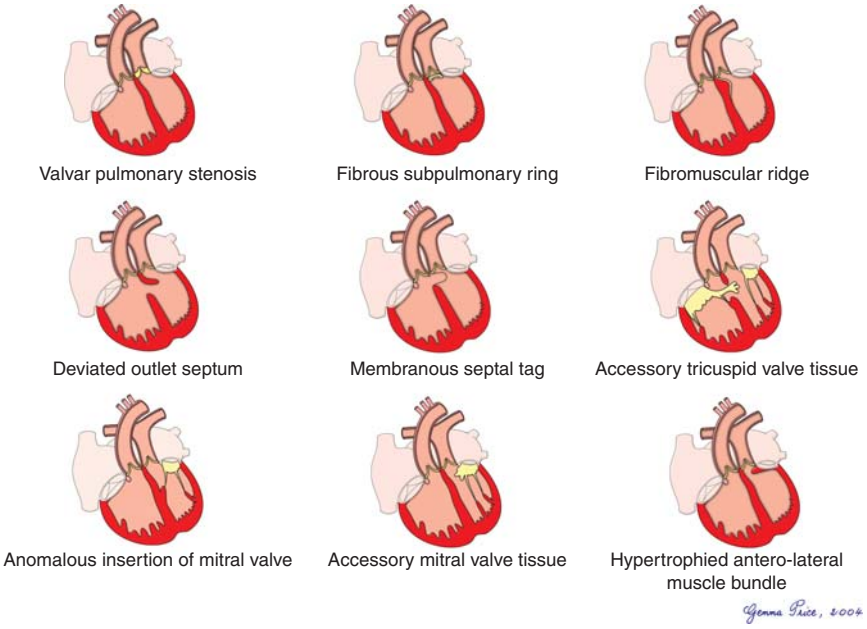

Figure 6.
The cartoon shows the various lesions that can produce obstruction of the left ventricular outflow tract. They produce subaortic obstruction when the ventriculo-arterial connections are concordant, but subpulmonary obstruction in the setting of transposition.

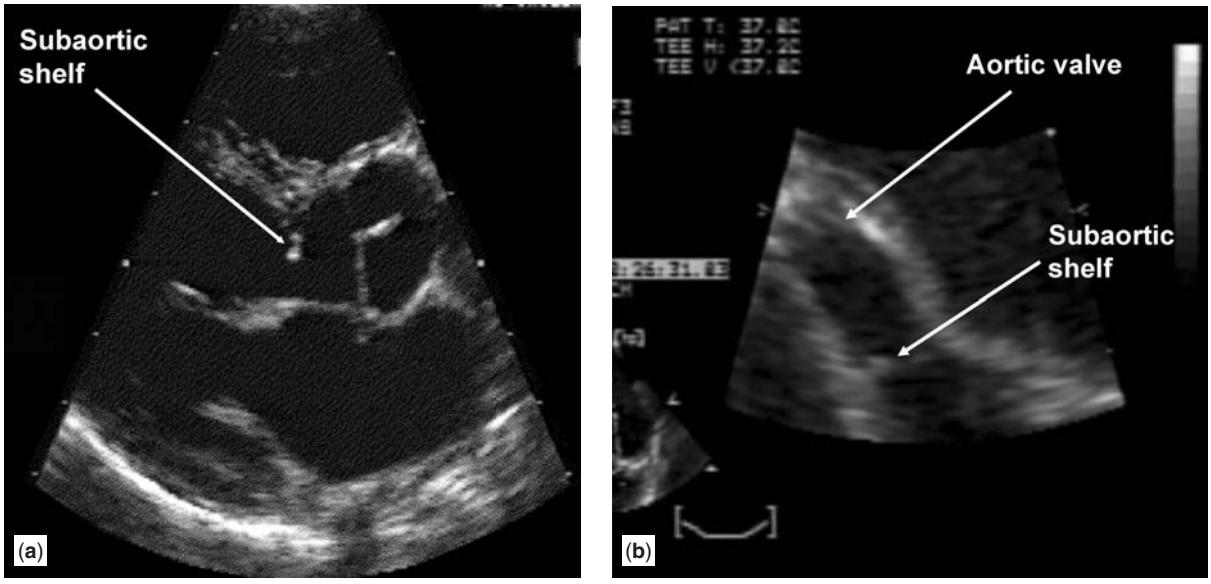

Figure 7.
*(**a**) taken in parasternal long axis view, shows a discrete subaortic shelf. (**b**) shows another shelf as seen in the apical 5-chamber view in a patient with Shone's complex.*

which may extend from the sinuses of Valsalva to the brachiocephalic artery.

Aortic regurgitation

Repair is now the first line of treatment taken by most surgeons dealing with the incompetent aortic valve. It is important, therefore, to understand the mechanism of aortic regurgitation so as to evaluate the adequacy of repair. Transoesophageal echocardiography allows better definition of the morphology, so intraoperative transesophageal echocardiogram is mandatory, since because the aorta is opened, and unlike the mitral valve, there is no good method of testing the aortic valve after repair. Appropriate surgical repair is tailored to the mechanism of valvar regurgitation. This can be a primary congenital anomaly of the valve itself, which can possess one, two, three or rarely four leaflets. Regurgitation can also be produced, however, by an aortic-left ventricular tunnel, or can be due to congenital unguarding of the aortic valve orifice. Aortic regurgitation may also be associated with other

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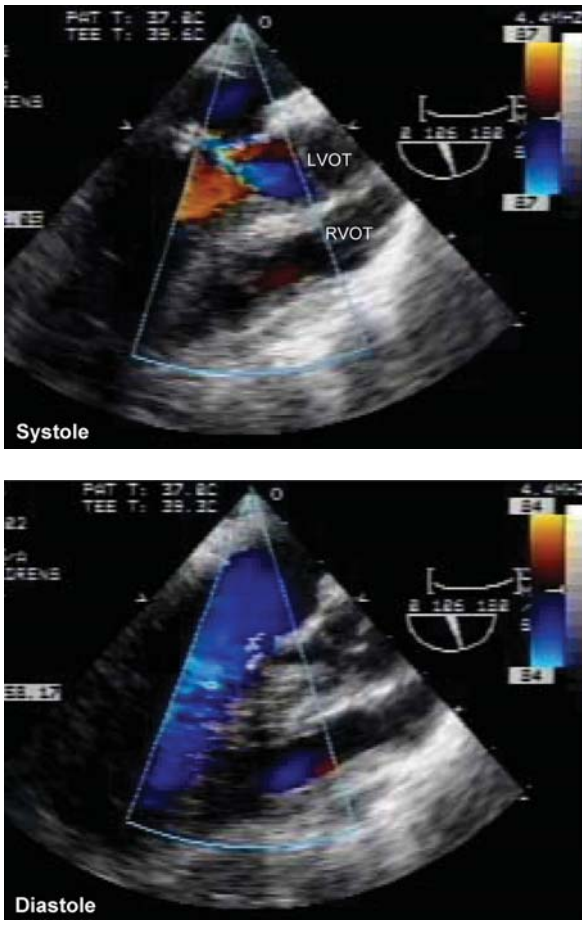

Figure 8.
The panels show a fibro-muscular subaortic tunnel, with systolic and diastolic frames showing the narrowed nature of the left ventricular outflow tract.

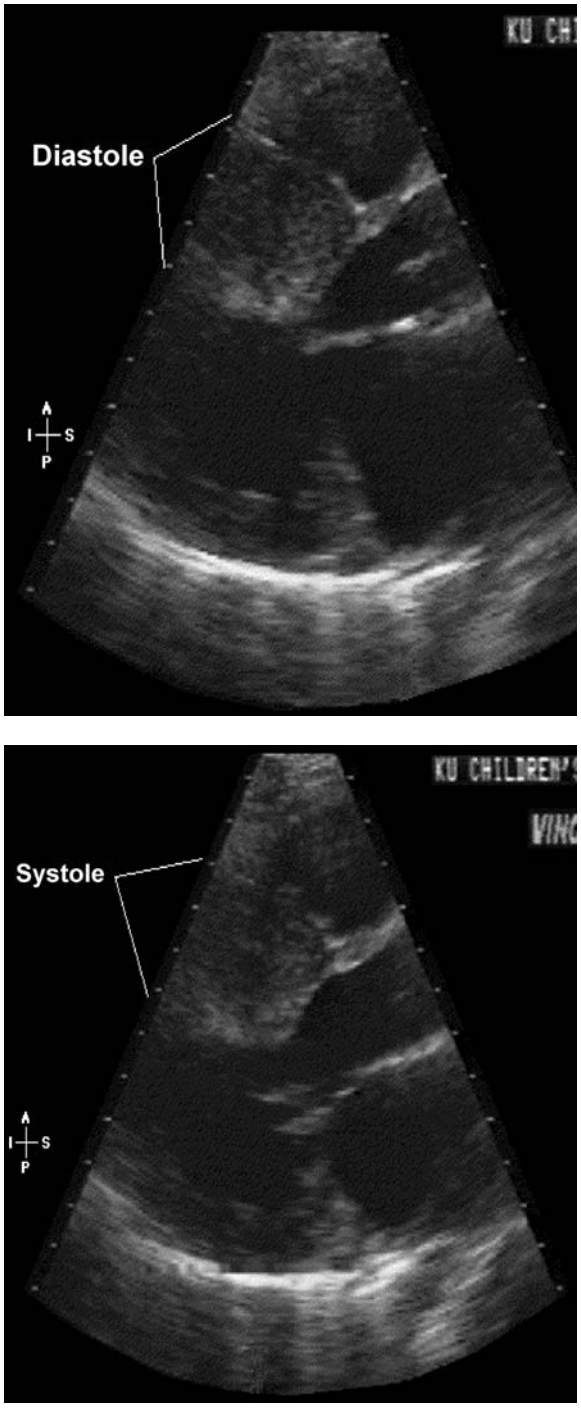

Figure 9.
The panels show systolic and diastolic frames of the left ventricular outflow in a patient with hypertrophic cardiomyopathy.

congenital malformations, such as perimembranous or doubly committed ventricular septal defects opening to the outlet of the right ventricle, subaortic stenosis, common arterial trunk, and tetralogy of Fallot.

Aortic regurgitation can also accompany dilation of the aortic root, as in patients with connective tissue disorders, and congenital aneurysms of the sinus of Valsalva (Fig. 11), as well as being a complication of an infectious process such as rheumatic fever or endocarditis. Intraoperative transoesophageal echocardiogram can identify the underlying surgical pathology, enabling the surgeon to tailor his or her repair to the particular lesion. Patients with annular dilation would require either commissural plication, or less commonly an encircling suture of the entire aortic circumference. Perforation of an aortic leaflet is repaired by simple closure with a patch. Repair of a prolapsing leaflet would consist of either triangular resection, or resuspension of the free edge. Commissural prolapse is a rare cause of aortic regurgitation caused by aortic dissection. Repair of this lesion is by resuspension of the commissures with pledgeted sutures that occlude the dissected space.

Ventriculo-arterial connections

As shown in the anatomic review,[1] there are four types of ventriculo-arterial connection, namely concordant,

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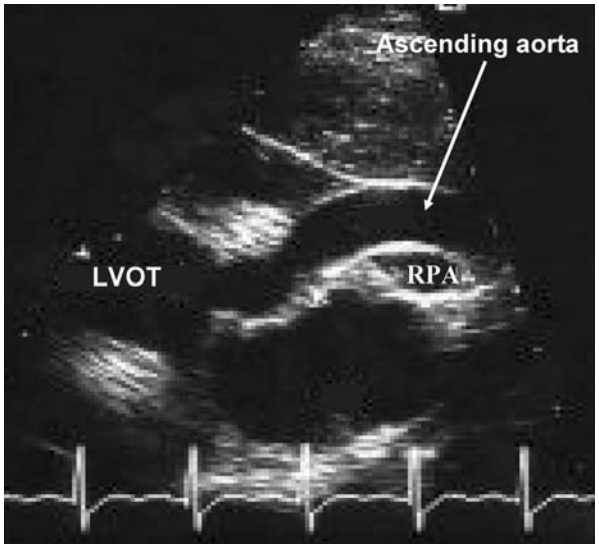

Figure 10.
This echocardiogram shows diffuse and uniform hypoplasia of the ascending aorta in a patient with Williams' syndrome.

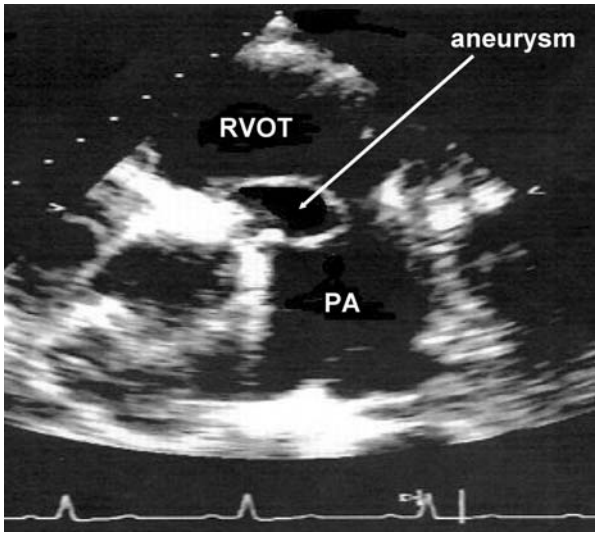

Figure 11.
In this patient, the echocardiogram shows an aneurysm of the sinus of Valsalva encroaching on the right ventricular outflow tract.

discordant, double outlet, or single outlet. The usual, or concordant, connections are seen when the aorta arises from the morphologically left ventricle, and the pulmonary trunk arises from the morphologically right ventricle. In this setting, the parasternal long axis and apical 5-chamber echocardiographic views reveal the fibrous continuity between the aortic leaflet of the mitral valve and the leaflets of the aortic valve, which is a key echocardiographic feature of normality.[4] Discordant connections exist when the aorta arises from the morphologically right ventricle, and the pulmonary trunk from the morphologically left ventricle. In most such instances, the arterial leaflet

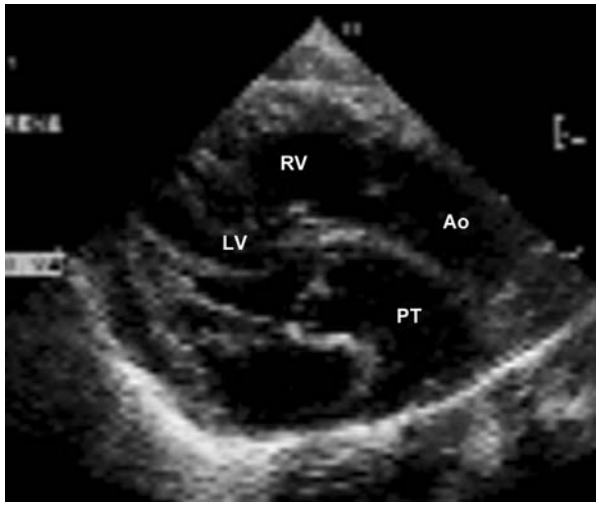

Figure 12.
The echocardiogram shows discordant ventriculo-arterial connections. Note the fibrous continuity between the leaflets of the pulmonary and mitral valves in the left ventricle.

of the mitral valve leaflet is in fibrous continuity with the leaflets of the pulmonary valve (Fig. 12). Double outlet connection exists when the greater parts of both ventriculo-arterial junctions are supported by the same ventricle. This ventricle may be of right, left, or very rarely indeterminate morphology. When categorizing the ventriculo-arterial connections, overriding arterial valves are assigned to the ventricle supporting the greater part of their circumference, as judged in the valvar short axis.

Single outlet is more complex, since it includes four types of connection. A common trunk is guarded by a common arterial valve, and gives rise directly to the coronary arteries, at least one pulmonary artery, and the majority of the systemic circulation. A solitary trunk is defined in the absence of the intrapericardial pulmonary arteries, since in this circumstance it is impossible to know, had they existed, whether the pulmonary arteries would have arisen from the heart itself or from an arterial trunk. As explained in the previous review, the other forms of single outlet are single aortic trunk with pulmonary atresia, and single pulmonary trunk with aortic atresia.

The left ventricular outflow tract in the setting of concordant atrioventricular but discordant ventriculo-arterial connections

Obstruction to the pulmonary outflow tract is common in the setting of transposition.[5] It can be produced at valvar, subvalvar, or supravalvar levels, and may occur in patients with an intact ventricular septum, or in association with a ventricular septal defect (Fig. 6). Dynamic subpulmonary stenosis is common in patients with intact ventricular septum, and also

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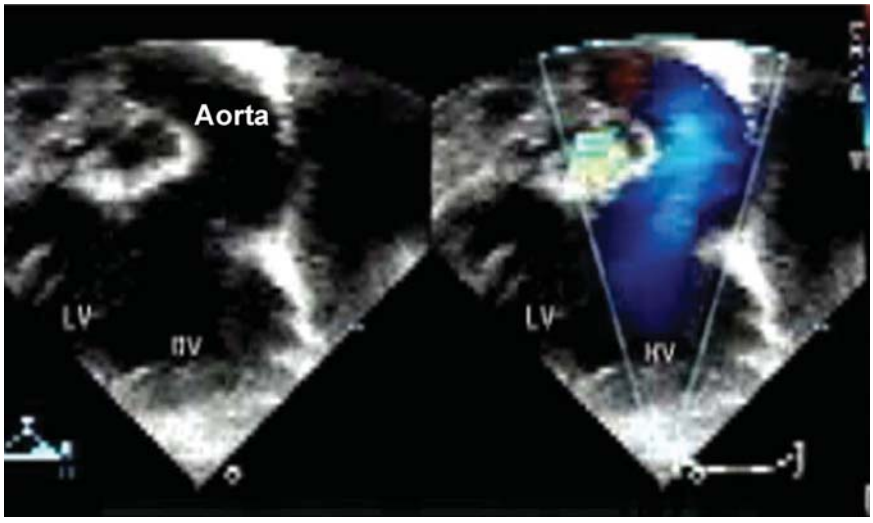

Figure 13.
In this patient with congenitally corrected transposition, the aorta is in left-sided and anterior position. It was necessary, however, to confirm the presence of discordant atrioventricular connections so as to establish the diagnosis.

after the Senning operation. The most common causes of fixed obstruction are a fibrous subpulmonary diaphragm, a fibromuscular ridge, and valvar stenosis due to a bifoliate pulmonary valve. All of these lesions are best seen echocardiographically in the parasternal long-axis, apical 5-chamber, or subcostal 4-chamber views. The transesophageal echocardiogram allows further definition of the mechanism of obstruction using multiple imaging planes.

The left ventricular outflow tract in the setting of discordant atrioventricular and ventriculo-arterial connections

The echocardiographic diagnosis of congenitally corrected transposition is made by demonstrating the abnormal connections between the atrial and ventricular chambers, in addition to the discordant ventriculo-arterial connections. In most cases, a clue is given to the likely diagnosis since, in most instances, the aorta is anterior and to the left of the pulmonary trunk (Fig. 13). This finding, however, provides only inferential evidence for the existence of corrected transposition, since the transposed aorta can also be left-sided in the setting of concordant atrioventricular connections, while the transposed aorta is usually right-sided when corrected transposition is seen in its mirror-imaged variant. It is essential, therefore, to establish the presence of discordant atrioventricular connections. This is best done using subcostal views, which reveal the connections between the cardiac chambers themselves and the arterial trunks when the transducer is tilted in different directions. In most instances, the aortic valve is supported by a complete muscular infundibulum, whereas the pulmonary valve is usually wedged deeply between the two atrioventricular valves. The aortic valve, therefore, is more superiorly located than the pulmonary valve. The parasternal echocardiographic view shows the fibrous continuity usually seen between the leaflets of the pulmonary and mitral valves. In the setting of deficient ventricular septation, it is also possible, in most instances, to demonstrate continuity between the leaflets of the pulmonary and tricuspid valves through the central fibrous body and the membranous septum. Obstruction to the left ventricular outflow tract occurs in about two-fifths of the patients with congenitally corrected transposition. It is most usually subvalvar, being caused either by a subvalvar diaphragmatic ring, or an aneurysm of adjacent fibrous tissue protruding in the outflow tract. These lesions are imaged well from the apical 4-chamber or subcostal views.

The left ventricular outflow tract in double outlet right ventricle

Echocardiography is now the technique used most frequently in evaluating the anatomy, haemodynamics, and function of the heart prior to and after surgical repair or palliation.[6,7] Currently, it is the most reliable means of establishing the diagnosis of double outlet from the right ventricle, albeit that this could well change as more units gain access to techniques producing tomographic images that permit the relationship of the circumferences of the arterial valve to be established relative to the crest of the muscular septum. It is still in the assignment of overriding arterial valves that most difficulties are encountered with making the diagnosis of double outlet, and three-dimensional echocardiography is also likely to provide significant added value when making such adjudications. The echocardiographer should be seeking to establish at least four features. The most important, but also the most difficult, is to be sure that both great arteries arise in their greater part from the morphologically

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right ventricle. The second point is to establish the morphology and site of the channel taking blood from the left ventricle. This channel, the interventricular communication, is usually also described as the ventricular septal defect. The third point is to establish the relationship of the aorta relative to the pulmonary trunk. Usually the aorta is to the right or anterior when there is usual atrial arrangement, but rarely the aorta can be left sided. It is important to establish the latter situation, since in this instance, the aortic position is not an indicator of congenitally corrected transposition (see above). The final feature is to establish the nature of the roof of the interventricular communication. When the outlet from the left ventricle leads directly into one or other of the arterial trunks, or both, it used to be argued that, to justify the diagnosis of double outlet right ventricle, the roof of the defect needed to be muscular, being composed of the subarterial infundibulum that separated the leaflets of the arterial valves from those of the mitral valve. It is now well-established, however, that both arterial trunks can arise exclusively from the right ventricle when there is fibrous continuity between the leaflets of the arterial and mitral valves. The significance of this point is that, with such fibrous continuity, the arterial valve will be much closer to the exit from the left ventricle, making it easier for the surgeon to reconnect one or other arterial outflow tract to the left ventricle.

The echocardiographic feature that points to the diagnosis of double outlet right ventricle is the finding of the muscular outlet septum exclusively within the right ventricle. This feature, of course, is not exclusive to double outlet, but is also seen in patients with tetralogy of Fallot, as well as in those with discordant ventriculo-arterial connections.[8] In both of these settings, there is a spectrum of malformations between the extremes of double outlet and one-to-one ventriculo-arterial connections. The finding of bilateral infundibulums can be helpful in diagnosing the double outlet connection, but as we have shown in the previous review, bilateral infundibulums can also be found in patients with transposition, and rarely in those with otherwise normal hearts. The key to diagnosis, therefore, is in determining the degree to which overriding arterial valves are connected within the morphologically right as opposed to the left ventricle, and this still remains difficult.

It is the location of the interventricular communication, or ventricular septal defect, that is usually used to form the basis of morphological and surgical classifications, although within this spectrum of variability, any position of the aorta can combine with any position of the pulmonary trunk, and with any location of the interventricular communication, thereby creating an infinite number of morphological possibilities. Certain combinations, nonetheless, stand out.

Subaortic defect. This is the most common sub-type, and is morphologically related to tetralogy of Fallot. The interventricular communication, which is essentially the primary outlet from the left ventricle, is located beneath the outflow tract supporting the aortic valve, and like in tetralogy of Fallot, the defect is positioned between the limbs of the septomarginal trabeculation. The leaflets of the aortic valve often override the crest of the ventricular septum. Although there can be discontinuity between the leaflets of the aortic valve and both atrioventricular valves, even in this setting there can be fibrous continuity between the mitral and tricuspid valvar leaflets. This latter feature is of surgical importance, since it makes the defect perimembranous, and indicates that the conduction tissue is closely related to the margin of the interventricular communication at its postero-inferior rim. Only very rarely in the setting of a subaortic defect is there discontinuity between the leaflets of the mitral and tricuspid valves.

Since the defect is the only outlet for the left ventricle, stenosis of the interventricular communication effectively produces subaortic obstruction, although there is, of course, free access to the aortic outflow tract from the right ventricle. In the majority of cases, however, the subaortic outflow tract is widely patent from both ventricles, and the outlet septum, which as already emphasised is an exclusively right ventricular structure, is deviated in antero-cephalad direction, often producing subpulmonary obstruction. Almost always the arterial trunks are normally related when the interventricular communication is subaortic, or side-by-side with the aorta to the right. As also emphasised above, in one rare variant the aorta can be anterior and to the left of the pulmonary trunk. The defect is almost always in subaortic position with this rare pattern, making it possible in most cases for the surgeon to connect the aorta directly to the left ventricle.

Subpulmonary defect. With this arrangement, the defect is again between the limbs of the septomarginal trabeculation, but in a subpulmonary location, giving so-called "Taussig-Bing heart". This pattern, the second most frequent, is typically associated with parallel alignment of the arterial trunks, the aorta being to the right and slightly or markedly anterior to the pulmonary trunk. It is the orientation of the outlet septum, an exclusively right ventricular structure, which is the central feature in a spectrum of malformation determined by the precise connection of the overriding pulmonary valve. Rightward deviation of the outlet septum produces subaortic obstruction, a frequent finding along with aortic coarctation.

Doubly-committed defect. This subgroup is also closely related to tetralogy of Fallot, but in the setting of absence of the muscular outlet septum, so that there

is fibrous continuity between the facing leaflets of the aortic and pulmonary valves. The large interventricular communication is located immediately beneath both the aortic and pulmonary valves, which have a marked tendency to override. Thus, the lesion is also related to double outlet left ventricle (see below).

The left ventricular outflow tract in double outlet left ventricle

The diagnosis of this lesion, a very rare anomaly, is made when both the aorta and the pulmonary trunk arise exclusively or predominantly from the morphologically left ventricle. Parasternal and subcostal imaging is usually required to make the diagnosis. In most instances, there is fibrous continuity between the arterial leaflets of the mitral valve and both the aortic and pulmonary valves. Pulmonary valvar and subvalvar stenosis is common, being found in about seven-tenths of the reported cases. Most patients with this ventriculo-arterial connection will lack either a complete subaortic or subpulmonary infundibulum.

Rarer anomalies involving the left ventricular outflow tract

Aorto-left ventricular tunnel. The defect usually involves the right aortic sinus of Valsalva, and is due to disruption of the semilunar attachment of the leaflet to the sinus. Although the tunnel is frequently said to extend through the muscular outlet septum, in reality it occupies the space between the free-standing subpulmonary infundibulum and the aortic root. It results in free regurgitation from the aorta to the left ventricle. There is usually a secondary aneurysmal dilation of the right aortic sinus of Valsalva. Eventually, the aortic root, ascending aorta, and the left ventricle all become enlarged. There may also be severe valvar regurgitation in addition to the regurgitation through the tunnel. The tunnel can, more rarely, involve the other sinuses of Valsalva, and on occasion can run from the aorta to the right ventricle.[9]

Aneurysm of sinus of Valsalva. This is a true aneurysm produced by a localized weakness in the wall of the sinus of Valsalva. It may be congenital or acquired, the latter usually being secondary to Marfan's syndrome, or endocarditis. The orifice of the aneurysm is just above the ventriculoaortic junction. The right aortic sinus is most commonly involved (Fig. 11). The aneurysm can rupture, or produce other complications such as thrombosis, obstruction of the right ventricular outflow tract, or compression of surrounding structures.

Aneurysm of the mitral-aortic intervalvar fibrous continuity. This lesion may also be congenital or acquired. When acquired, it can be secondary to infection or trauma, or appear after surgical insertions of homografts or prosthetic valves. The mouth of the defect is proximal to the attachment of the leaflets of the aortic valve (Fig. 14).

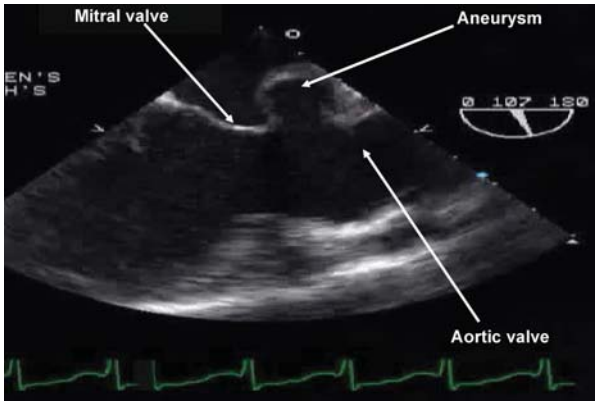

Figure 14.
The panels show an aneurysm involving the area of fibrous continuity between the leaflets of the mitral and aortic valves. Note that the mouth of the aneurysm is below the attachments of the aortic valvar leaflets.

Conclusion

Echocardiography is an effective and extremely useful method for diagnosing the malformations, often complex, which can afflict the left ventricular outflow tract. The test accurately identifies the anatomical variables, and guides the choice of the most appropriate surgical approach.

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