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Original Article

Echocardiography of the tricuspid and pulmonary valve in children*

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Abstract Diseases of the tricuspid and pulmonary valve are common in childhood. These include congenital anomalies, acquired lesions, and secondary valve compromise due to left heart disease. A comprehensive and methodical approach to the echocardiographic assessment of these diseases of the tricuspid and pulmonary valve is necessary for best care of children with these conditions.

Keywords: Tricuspid insufficiency; tricuspid stenosis; pulmonary insufficiency; pulmonary stenosis; echocardiography

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Diseases of the tricuspid and pulmonary valve are common in childhood. These include congenital anomalies, acquired lesions, and secondary valve compromise due to left heart disease. A comprehensive and methodical approach to the echocardiographic assessment of these diseases of the tricuspid and pulmonary valve is necessary for the best care of children with these conditions.

Tricuspid valve disease

The tricuspid valve plays an important role in normal newborn and paediatric cardiac physiology. Primary abnormalities of the tricuspid valve may contribute significantly to haemodynamic impairment. In addition, the tricuspid valve is often impacted secondarily from other congenital heart defects and changes in myocardial performance.

The tricuspid valve has several anatomic features including leaflet tissues, chordae tendineae, supporting annular ring, as well as a subvalvar apparatus that includes the papillary muscles. It is the cardiac valve with the largest orifice. The tricuspid valve has three leaflets – anterior, posterior, and septal – which are attached to a fibrous annulus and are unequal in size. The largest leaflet is the anterior leaflet, which extends from the anterolateral wall posteriorly to the infundibulum region anteriorly. The septal leaflet extends from the interventricular septum to the infundibulum and, as the name suggests, has numerous attachments to the ventricular septum. The posterior leaflet attaches along the posterior margin of the septum to the anterolateral wall. The tricuspid annulus is not a planar structure. Rather, it has an elliptical shape resembling a saddle, which is important both in the function and in the consideration of anatomic repair.

A number of imaging planes can be valuable in assessing tricuspid valve anatomy and function. Subcostal, coronal, and sagittal imaging planes demonstrate en face views. These views can identify areas of coaptation or lack thereof. In addition,

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subcostal views allow one to trace the attachments of the valve to the subvalvar apparatus. Similarly, parasternal short-axis imaging can demonstrate the anatomy of the tricuspid valve leaflets. Parasternal long-axis imaging conversely lends itself to assessment of tricuspid valve mobility and is ideally suited to assess flow across the valve and insufficiency of the valve. Apical imaging is well suited to assess the dimensions of the tricuspid valve as well as obtain meaningful Doppler data. Measurement of the tricuspid valve annulus is critical to decision making in a number of congenital anomalies. Such measurements are obtained from apical imaging and then expressed as a Z score.[1] The apical image often lines up parallel with the inflow across the tricuspid valve and is suitable for a pulse wave Doppler interrogation of tricuspid valve inflow.

Among the more valuable uses of echocardiography and the assessment of the tricuspid valve is determination of the degree of tricuspid regurgitation as well as the haemodynamic profile of the regurgitant tricuspid jet. In general, inflow across the tricuspid valve is largely dependent on a number of physiologic parameters. It is particularly influenced by intrathoracic pressure. As a result, when performing Doppler interrogation across the tricuspid valve it is essential to sample throughout the cardiac and respiratory cycle. Failure to account for this can sometimes lead to overestimation or underestimation of calculated tricuspid valve inflow gradients. The ideal method for determining tricuspid inflow is the use of pulse wave velocity with interrogation at the leaflet tips of the tricuspid valve.[2]

Interrogation of tricuspid regurgitation is important in a number of clinical decisions, including the assessment of the severity of regurgitation. The quantification of tricuspid regurgitation using flow Doppler techniques has been widely described. The American Society of Echocardiography has established guidelines regarding the evaluation of native valvar regurgitation, including that of the tricuspid valve.[3] Interrogation of a tricuspid valve should include colour flow Doppler, which provides a visualisation of the origin of the regurgitant jet at its width (vena contracta), the spatial orientation of the regurgitant jet area in the atrium, and the flow convergence into the regurgitant orifice. Using these principles rather than the regurgitant jet area alone improves the accuracy of colour flow Doppler for assessing the severity of tricuspid valve regurgitation. Conversely, the size of the regurgitant jet by colour flow Doppler may have limitations related to gain, Nyquist limit, size, and depth of the image sector. Although a comprehensive review of the methodologies for colour flow Doppler interrogation of valve regurgitation is beyond the scope of this paper, assessments of the regurgitant jet area, the vena contracta, and the proximal isovelocity surface area are of value. Pulse wave Doppler interrogation for quantitation is less valuable given a number of limitations related to the principles of velocity time integral. This is generally not used regularly in the assessment of paediatric tricuspid valve disease. Pulse wave Doppler and continuous wave Doppler are used frequently to determine the peak velocity of the tricuspid regurgitation. This has been well established to correlate with catheter measures of right ventricular and systolic pressure. It is included in a majority of comprehensive paediatric echocardiography exams.

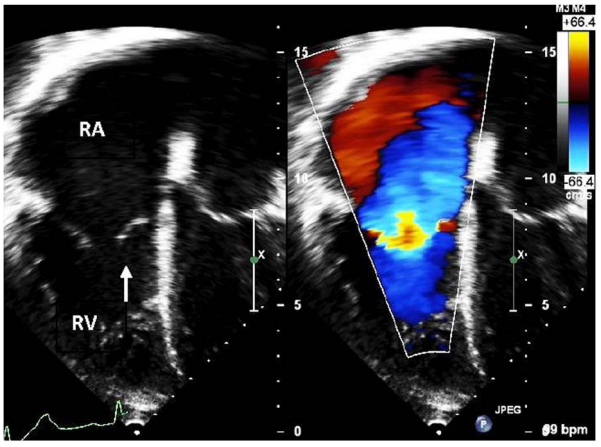

Figure 1.
Apical image demonstrating severe tricuspid regurgitation secondary to tricuspid valve injury from prior endomyocardial biopsy. Disruption of the chordae tendineae of the tricuspid valve leaflet are shown. RA = right atrium; RV = right ventricle.

In addition to physiologic interpretation of tricuspid regurgitation, echocardiography can identify the underlying cause of valve regurgitation. These include acquired conditions such as infective endocarditis and rheumatic fever.[4] Iatrogenic causes should also be considered; these would include prior cardiac surgery, biopsies, or catheter placements (Fig 1). Finally, a number of the congenital heart lesions described will lead to tricuspid valve insufficiency. One of the more common causes of tricuspid regurgitation is tricuspid annular dilatation. This may be seen in the setting of chronic right ventricular dilation, or in the setting of elevated right heart pressure secondary to left heart disease.

Specific lesions

Ebstein's anomaly of the tricuspid valve is characterised by apical displacement of the septal leaf of the tricuspid valve. There is tethering of the leaflet of the ventricular septum, and the portion of the right ventricle above the valve annulus is atrialised and thin-walled. The echocardiographic assessment of Ebstein's anomaly focusses on a number of key

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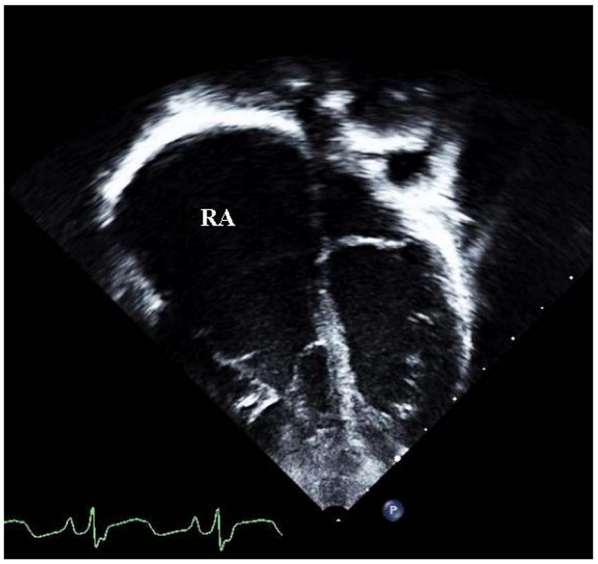

Figure 2.
Apical image demonstrating classic features of Ebstein's anomaly with tethering of the septal leaflet and downward displacement of the zone of apposition of the tricuspid valve leaflets. RA = right atrium.

features.[5,6] The apical four-chamber view demonstrates the displacement of the septal leaflet of the tricuspid valve (Fig 2); complementary short-axis views of the heart will demonstrate elongation of the anterior and posterior tricuspid valve leaflet. It is important to note that the affected orifice of the tricuspid valve is apically or infundibularly displaced (Fig 3). Short-axis views, therefore, need to sweep through the entire right ventricle to identify the affected tricuspid valve orifice and the origin of tricuspid valve regurgitation. In terms of tricuspid valve morphology it is important to define the following:

1. Severity of septal leaflet offset – best determined in apical four-chamber view.
2. Severity of posterior leaflet offset – best determined in apical four-chamber view.
3. Anterior leaflet size and mobility – determined in parasternal long-axis view, parasternal short-axis view, and apical four-chamber view.

Echocardiography can be useful in assessing the likelihood of effective repair of the tricuspid valve. A paper by Brown et al[7] in 2008 demonstrated that, with the use of several echocardiographic features, one could have a sensitivity of 59% and a specificity of 92% for achieving effective repair of the tricuspid valve. In addition, it can be helpful to assess the right ventricular size. This can be done qualitatively for a parasternal short-axis view, or apical four-chamber view, as well as subcostal imaging. The diameter of the right ventricular base in apical four-chamber view can be helpful. This will allow assessment of

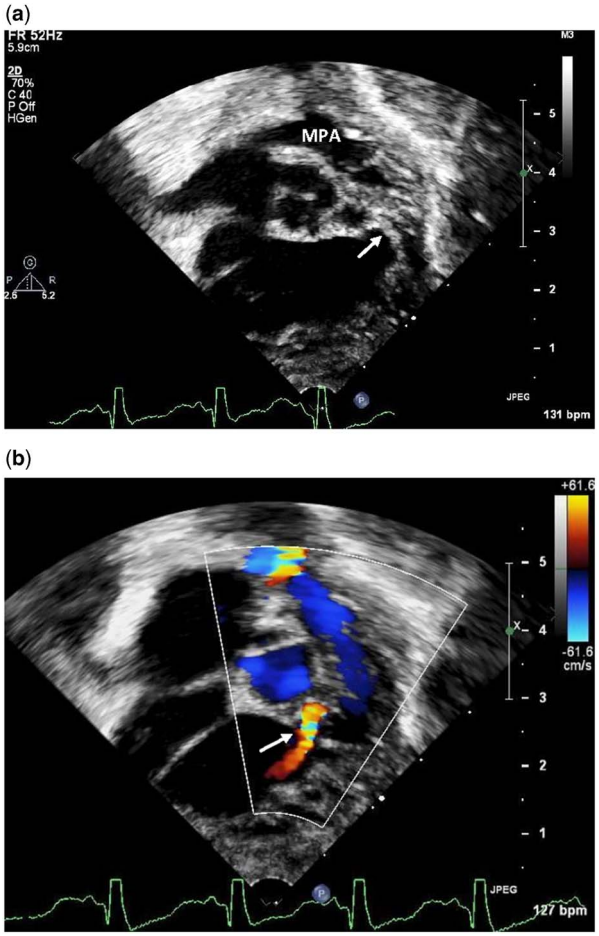

Figure 3.
*(**a**) Subcostal RAO image demonstrating apical displacement of the tricuspid valve in Ebstein's anomaly with the effected orifice of the tricuspid valve being into the right ventricular outflow tract. (**b**) Coronal subcostal image demonstrating tricuspid valve insufficiency oriented towards the transducer, highlighting the displacement of the tricuspid valve towards the right ventricular outflow tract. MPA = main pulmonary artery = pulmonary trunk.*

the degree of atrialisation of the right ventricle. Moreover, it is important to assess right ventricular function in this setting. This is often best determined in a parasternal long-axis outflow view. Tricuspid valve Doppler inflow imaging and tissue Doppler imaging can also be quite helpful. The pulmonary artery may be impacted in Ebstein's anomaly. In some cases, there is functional atresia of the pulmonary valve and very little antegrade blood is ejected by the right ventricle. Therefore, it is important to measure the pulmonary valve annulus size, the main pulmonary diameter, and Doppler interrogation for pulmonary stenosis. One other feature that is important in the echocardiographic assessment of Ebstein's anomaly is determination of the atrial shunt. In some patients with more severe forms of Ebstein's anomaly there will be bi-directional atrial shunt or predominant right to left atrial shunt.[8] This

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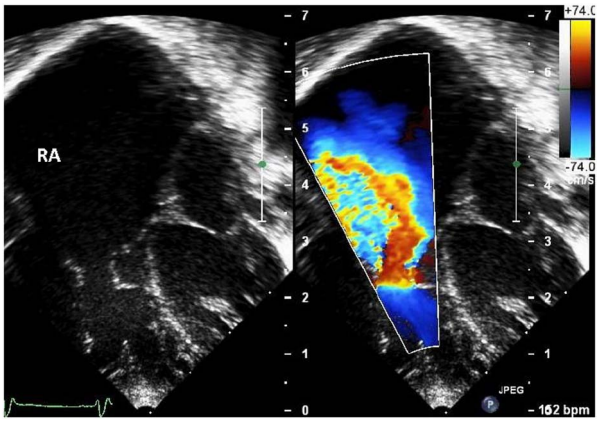

Figure 4.
Apical image demonstrating congenital dysplasia of a tricuspid valve with an area of non-coaptation and a broad jet of severe tricuspid regurgitation. RA = right atrium; RV = right ventricle.

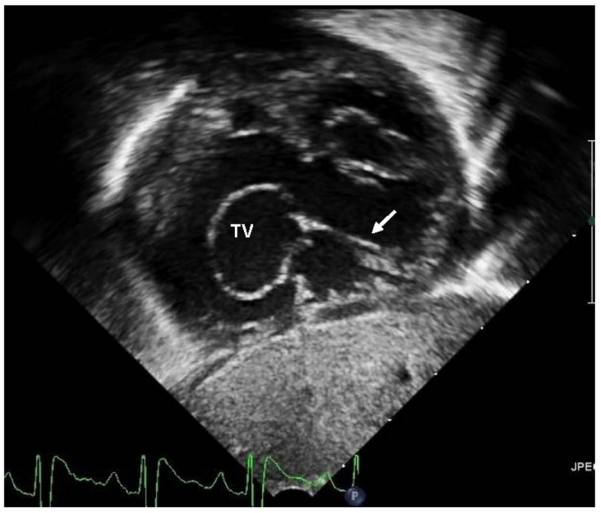

Figure 5.
Subcostal sagittal view demonstrating tricuspid valve straddle. The chordae tendineae of the tricuspid valve insert into the left ventricle papillary muscle towards the base of the left ventricle. TV = tricuspid valve.

can ideally be done by subcostal coronal imaging. The shunt direction can be determined by colour flow Doppler. When colour flow Doppler is not confirmatory, saline contrast injection with a Valsalva manoeuver can be incorporated. It is important to recognise that left ventricular function may also be compromised in Ebstein's anomaly and this should be assessed by echocardiography.

Isolated cases of tricuspid valve insufficiency unrelated to Ebstein's anomaly are occasionally seen in childhood. A variety of anatomic abnormalities can lead to primary tricuspid valve regurgitation. These anatomic abnormalities include shortened chordae tendineae and hypoplastic or absent papillary muscles. The echocardiographic assessment of these lesions is relatively similar to the echocardiographic assessment of Ebstein's anomaly (Fig 4).

A rare anomaly of the tricuspid valve is the so-called unguarded tricuspid valve orifice. This results in a very severe form of tricuspid valve regurgitation.[9] In this scenario, the tricuspid valve leaflets are never fully formed in foetal development. This results in unimpeded reverse flow from the right ventricle into the right atrium in the systole. On colour flow Doppler interrogation, one notes a large broad jet of tricuspid regurgitation. In the short axis, one can detect the hypoplastic or absent tricuspid valve leaflets.[10] Typically, this lesion is associated with rather significant right atrial and right ventricular dilatation.

Primary tricuspid stenosis of the tricuspid valve is relatively uncommon. Tricuspid valve hypoplasia may be seen in a number of other conditions such as pulmonary atresia and intact ventricular septum. Tricuspid stenosis can also be iatrogenic or a result of previous surgical interventions, including attempts to repair the tricuspid valve or other surgical procedures. In adults, tricuspid valve disease is predominantly related to rheumatic heart disease rather than being a congenital anomaly. The most common setting for tricuspid valve stenosis in the current era is as a complication of previous tricuspid valve repair. Most commonly, tissue valves placed in the tricuspid position for severe tricuspid valve disease have become calcified over time. Echocardiography can help determine the inflow gradient across the tricuspid valve. In addition, echocardiography can identify calcification of the tricuspid valve leaflets.[11]

Another rare anomaly is double outlet of the right atrium. In this case there are two atrioventricular valves that both enter into the right ventricle.[12] This can be considered an analogue of a double orifice mitral valve. This is best imaged in a parasternal short-axis view or in a subcostal sagittal view.[13]

It is key to always evaluate for abnormal attachments of the tricuspid valve leaflet apparatus, especially in the setting of a ventricular septal defect. Anomalous tricuspid valve attachments are most commonly associated with inlet ventricular septal defects. Straddling of the tricuspid valve is characterised by valvar attachments to both the left and the right ventricles.[14] In contrast, overriding of the tricuspid valve occurs when the annulus is committed to both ventricles (Fig 5). Straddling must be identified before operative interventions to allow the surgeon to plan for repair of abnormal valvar attachments, or defer closing the ventricular septal defect if valve repair is not deemed feasible. Two-dimensional sweeps (loops) in multiple views are crucial to identifying straddling/overriding as a single 2D frame in one plane may not demonstrate abnormal tricuspid valve attachments.[15]

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Pulmonary valve

Congenital anomalies of the pulmonary valve are relatively common. These can be characterised by hypoplasia of the pulmonary valve annulus, or a normal-sized pulmonary valve annulus, but limited excursion of the pulmonary valve leaflets. The pulmonary valve is characterised by three relatively equally sized cusps. Pulmonary valve leaflets are thinner because of the lower pressures of the right heart system. A number of imaging planes can provide beautiful anatomic images of the pulmonary valve. Given the position of the pulmonary valve within a normal heart, parasternal imaging is often the most valuable. Parasternal long-axis view demonstrates the excursion of the pulmonary valve leaflets. In a normal pulmonary valve, leaflet edges extend in systole so that they nearly abut the walls of the main pulmonary artery. In the short-axis view, it may be difficult to obtain an en face view of the pulmonary valve. In the normal heart, rotation of the transducer from the parasternal long-axis to short-axis view tends to foreshorten the images of the pulmonary valve. Conversely, in certain disease states such as pulmonary hypertension or dilatation of the right ventricle, the angle of the pulmonary valve is moved, and in such cases a clear en face view of the pulmonary valve leaflet can be obtained.

Doppler interrogation of the pulmonary valve can be obtained in a number of imaging planes.[16] Subcostal sagittal generally affords the best alignment of the ultrasound beam to the outflow of the pulmonary valve. From this imaging plane, one can typically obtain a very reliable Doppler tracing that can be used to assess the flow velocity across the pulmonary valve. In this view, pulse wave Doppler across the right ventricular outflow tract and then continuous wave Doppler for high-velocity signals can be used to determine a pulmonary valve gradient. Although some controversy exists as to how best to report the pulmonary valve Doppler velocity, particularly in cases of pulmonary stenosis, recent studies have suggested that a mean Doppler gradient corresponds most closely to what is obtained by direct measurements in the cardiac catheterisation laboratory.[17] Pulmonary valve stenosis can be characterised by the movement of the pulmonary valve leaflets, as well as their thickness. This is best assessed in a parasternal long-axis view. In some disease states, the term dysplasia of the pulmonary valve has been utilised. The hallmark of this condition would be markedly thickened pulmonary valve leaflets with rolled or myxomatous edges of the valve leaflets (Fig 6).

Pulmonary valve stenosis lends itself quite well to transcatheter therapy. The current standard is balloon dilation for isolated pulmonary valve stenosis.

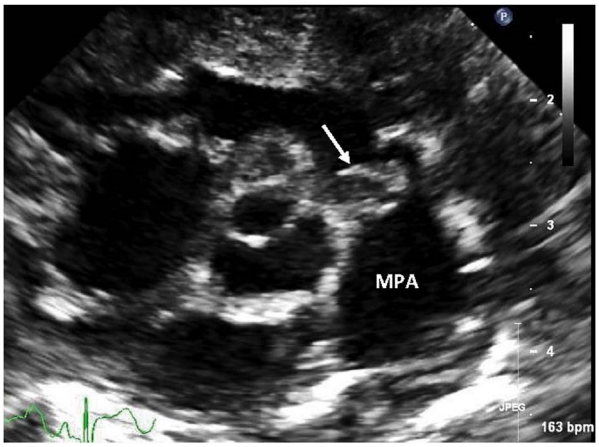

Figure 6.
Parasternal short-access imaging demonstrating severely dysplastic pulmonary valve leaflets in the setting of pulmonary stenosis with some dilatation of the main pulmonary artery. MPA = main pulmonary artery = pulmonary trunk.

In order to effectively carry out this procedure, it is important to understand a couple of anatomic features. Primarily, determination of the pulmonary valve annulus is critical to balloon sizing for this procedure. This measurement is most typically made in parasternal long-axis and parasternal short-axis imaging planes. In addition, it is crucial to exclude the co-existence of supravalvar or pulmonary stenosis.[18] This lesion often occurs in settings of genetic syndromes. The area of colour flow Doppler flow acceleration may make it difficult to distinguish from primary valve pulmonary stenosis. Careful two-dimensional imaging of the supravalvar region will help distinguish doming of the pulmonary valve leaflets from an impingement on the supravalvar region as one might see with supravalvar pulmonary stenosis. One can also note thickened walls of the main pulmonary artery in the setting of supravalvar pulmonary stenosis. It is essential to carefully interrogate the branch pulmonary arteries in this setting, and significant branch pulmonary artery disease may co-exist with supravalvar pulmonary stenosis.

In the setting of pulmonary valve stenosis, several other echocardiographic features can be quite valuable in the care of the child. Although Doppler interrogation will provide a measure of the pulmonary valve gradient, it is helpful to obtain complementary data regarding right heart pressure. Not uncommonly, in the setting of pulmonary valve stenosis there will be some degree of tricuspid regurgitation. Continuous wave Doppler interrogation of the tricuspid regurgitation should demonstrate elevation in right heart pressure consistent with Doppler values obtained across the pulmonary valve. In addition, in the setting of pulmonary stenosis, it is important to consider a number of other features. In some cases, pulmonary

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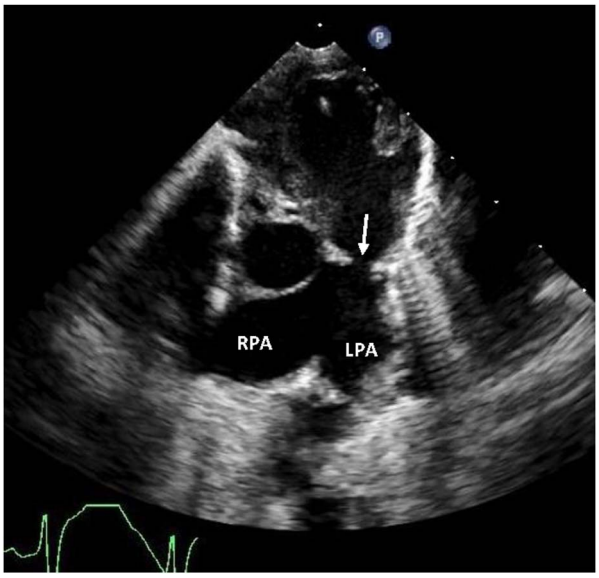

Figure 7.
Parasternal short-access image demonstrating an isolated absent pulmonary valve. In addition, one notes significant dilation of the branch pulmonary arteries. RPA = right pulmonary artery; LPA = left pulmonary artery.

valve stenosis is associated with varying degrees of right heart hypoplasia. Although standard two-dimensional echocardiography has limitations in terms of quantifying the right ventricular volume, assessment of the tricuspid valve annulus is thought to be a valuable surrogate for right heart size. In congenital heart conditions in which there is no ventricular septal defect, the tricuspid valve annulus dimension expressed as a Z score can often be considered a relatively reliable marker of overall right ventricular volume. When there is concern about right ventricular hypoplasia, measurement of the tricuspid valve dimension typically in an apical four-chamber view is essential.

Absent pulmonary valve most commonly occurs in the setting of tetralogy of Fallot. There have been some cases of isolated absent pulmonary valve. In this case, the pulmonary valve leaflets are present but often rudimentary (Fig 7).[19] The leaflet edges are rolled and the pulmonary valve annulus may be somewhat small relative to the body size. The colour flow Doppler feature suggests severe pulmonary regurgitation and sometimes co-existing mild pulmonary stenosis.[20]

In the quantification of pulmonary valve regurgitation, as is the case with the tricuspid valve, analysis of the three components of the regurgitant jet with colour flow Doppler are important. These three components are flow convergence zone, vena contracta, and jet turbulence. Together, these three components allow for an accurate assessment of the degree of pulmonary regurgitation. It is important to know that trace amounts of pulmonary regurgitation have been reported in up to 78% of the healthy population.[3]

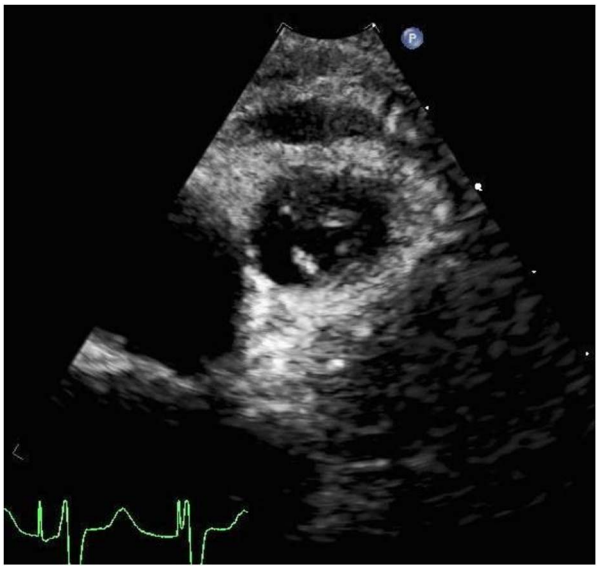

Figure 8.
Parasternal high short-access view demonstrating a bicuspid pulmonary valve.

The mechanisms of pulmonary regurgitation are variable. There may be congenital anomalies like quadricuspid or bicuspid valves (Fig 8). The most common cause of significant pulmonary regurgitation in the paediatric population tends to be common after repair of tetralogy of Fallot. There is no clinically accepted method for quantifying pulmonary regurgitation using continuous wave Doppler. When severe pulmonary regurgitation is evident, it is important to assess the right heart function and size. There have been a number of techniques used to assess right ventricular volume that are beyond the scope of this paper. Most commonly, centres use a semi-quantitative approach to describe the degree of right ventricular dilation that occurs in the setting of pulmonary regurgitation. It should be recognised that, in the current era, multimodality imaging offers the most comprehensive assessment of valve disease, especially right heart valve disease. As such, echocardiography should be paired with other imaging strategies such as cardiac MRI to improve clinical decision making.

In summary, echocardiography is invaluable in the assessment of tricuspid and pulmonary valve disease. The echocardiographic exam should recognise the benefit of complementary orthogonal imaging planes to gather clinically relevant data. The examiner also needs to know what data elements are essential for guided medical, transcatheter, or surgical therapy.

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None.

Ethical Standards

The authors assert that all procedures contributing to this work comply with the ethical standards of the relevant national guidelines.

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