# Simplifying ultrasound assessment of the fetal heart: incorporating the complete Three Vessel View into routine screening

# Abstract

*Introduction:* As our experience and ultrasound resolution have improved significantly in last 30 years it is possible to detect most of the major cardiac abnormalities prenatally with high degree of accuracy.

*Method:* Current screening techniques have poor detection rate for congenital heart diseases (CHD) and nearly half of the major cardiac abnormalities mainly of Great Arteries goes unrecognised. A high detection rate for major CHDs can be achieved in a screening setting by improving techniques of routine screening in the low risk population.

*Conclusion:* This article summarises the alternative strategies to examine the outflow tracts and Great Arteries, its correct interpretation and examples of abnormal views.

Keywords: cardiac defects, fetal echocardiography, prenatal diagnosis, ultrasound



Figure 1: Normal Three-Vessel View.

#### Introduction

Structural cardiac defects are the commonest birth defects, affecting 3–8/1,000 live births and comprising 28% of all birth defects in Australia.<sup>1-3</sup> Congenital heart disease (CHD) is six times more common than chromosomal abnormality and is associated with high rates of morbidity and mortality, accounting for 20% of stillbirths and 30% of neonatal deaths. Nearly 40% of babies with CHD require intervention after birth.<sup>4–7</sup> Ultrasound evaluation of the structure of the fetal heart has been possible for nearly 30 years. Historically, fetal echocardiography was performed by the paediatric cardiologist and was limited to high risk groups based on factors derived from maternal history: a family history of congenital heart disease (CHD), a history of chronic maternal disease (e.g. Diabetes Mellitus) or exposure to teratogenic medications (e.g. antiepileptic medicines and lithium). This policy has been shown to place 2–5% of women in a 'high-risk' group but detects only 5–10% of all major cardiac defects. Most cases of CHD occur in the low risk population, and can therefore only be detected through routine screening.<sup>8,9</sup> An effective strategy for prenatal screening allows early detection, improves antenatal, intrapartum and postnatal management, and reduces morbidity in survivors.<sup>10–15</sup>

Ultrasound is highly specific and sensitive for detection of cardiac abnormalities. As our experience and the resolution of ultrasound technology have continued to improve, it is now possible to detect most major cardiac abnormalities prenatally with a high degree of accuracy.16 Although 60% of major defects can be detected using the 4-chamber view alone, several major CHDs (transposition of the Great Arteries, tetralogy of Fallot, truncus arteriosus, and double-outlet right ventricle) are typically missed unless views of the outflow tracts are included. Inclusion of these views in routine screening potentially allows 80% of major structural cardiac abnormalities to be defined prenatally.<sup>17-21</sup> Several groups have, however, found that it is difficult to achieve these detection levels in routine clinical practice and there is significant variation in the effectiveness

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of screening even across centers within one country.<sup>22</sup>

While Australia has a strong obstetric screening program -98% of eligible women have a routine second trimester ultrasound scan - the detection rate for major cardiac abnormalities is poor. Population based studies evaluating the effectiveness of screening programs for the detection of cardiac abnormalities in unselected populations from South Australia, Victoria and NSW have concluded that the detection rate, particularly of major outflow tract abnormalities, is poor and is in the range of only 15-22%.<sup>23-25</sup> Koo, et al. in a population analysis from South Australia reported that routine obstetric ultrasound infrequently detects CHD. Only 22.5% of cases were referred for fetal echocardiography and prenatal detection of Transposition of the Great Arteries (TGA) and Tetralogy of Fallot occurred in only 15% and 25% respectively.23 Victorian data also show wide variation in antenatal detection rates ranging from 84.6% for hypoplastic left heart syndrome to 17% for TGA (17%).<sup>24</sup> Jaeggi, et al. reported that in NSW routine obstetric ultrasound failed to identify most duct dependent lesions. Abnormalities of the Great Arteries were identified in only 6.7% of cases.<sup>25</sup> This differs from data recently reported by Sholler, et al. who found that 44% of infants requiring cardiac surgery in first year of life had an antenatal diagnosis; but its important to recognise that this population has some bias as it does not include infants that died before surgery could be performed.<sup>26</sup> Nevertheless, the study confirmed that the majority of major cardiac abnormalities go unrecognised. It is likely that improvement in the detection rate will only be made by improving techniques of routine screening in the low risk population.

The current morphology screening protocol, described by the Australasian Society of Ultrasound in Medicine and adopted by most Australian centers, includes assessment of both the four chamber view and the outflow tracts and the long axis view of both ductal and aortic arches. The ductal arch is formed by the ductus arteriosus as it travels from its origin at the pulmonary artery to the point of entry into the descending aorta. Its distinguishing features include a relatively flat "hockey stick" shape and the fact that it does not give off any branches. The aortic arch on the other hand is more



Figure 3: Arch view (Color Doppler).

rounded (like a candy cane) and gives off branch vessels from its superior aspect (brachiocephalic, left common carotid and left subclavian arteries). Long axis views for the ductal and aortic arches were adopted from pediatric cardiology. The continuity of the arch may be confirmed if ideal views can be obtained in this plane, but both arches cannot be visualised simultaneously so discordance in size cannot be assessed and interruption of the aortic arch may be misinterpreted. Long axis views are difficult to acquire and interpret in normal fetuses and are more complex in the presence of an abnormality of the Great Arteries.

An alternative strategy for examination of the outflow tracts and Great Arteries is based on assessment of a series of axial sections, running from the four chamber view through the left and right outflow tracts through a view of the three vessels and finally of the transverse aortic and ductal arch. Imaging both the three vessels and the transverse arch are necessary to complete this assessment. These five essential views in transverse section are required for routine screening of CHD. These views are very reliable and easy to perform and interpret with only a short learning curve.<sup>27</sup>

#### The Three Vessel View: 2D and colour Doppler assessment

The Three-Vessel view (3VV) is above the four-chamber view (4chV) and is obtained by simply moving the ultrasound probe cranially from the 4chV (Figure 1). This view includes, from left to right: the main pulmonary trunk in direct communication with the ductus arteriosus, a transverse section of the aortic arch and the superior vena cava (SVC). The lumen of the aorta and pulmonary trunk are similar in size, though the pulmonary artery tends to be slightly larger and the SVC is the smallest vessel.<sup>28</sup> The cross section of trachea is also visible in front of spine. The head and neck vessels from the aortic arch arise some distance from the aortic valve and are not readily visible in these transverse views. The pulmonary artery branches laterally soon after its origin and this feature should be positively identified in transverse views.

Moving the probe further cranially images the transverse aortic arch, and the ductus arteriosus (Figure 2). The aortic arch crosses the midline from right to left. In this view the size



Figure 4: Ventricular disproportion.

and position of the transverse aortic arch and the arterial duct can be compared. Usually the duct is slightly smaller than the aortic arch at mid trimester ultrasound. The two vessels lie close together in this view and form a V shape before they join in front of spine. On color Doppler imaging both Great Arteries shows forward unaliased flow. The cross section of trachea is also seen in this section located anterior to the spine and to the right of both the duct and the aorta (Figure 3). This was defined as the three-vessel tracheal view by Yagel (2002) and more appropriate term for this view is a transverse arch views.<sup>27</sup>

As the normal three vessels are arranged in a straight line in decreasing order of diameter, subtle changes are readily appreciated. In addition to the vessel arrangement and alignment, other aspects such as size, abnormal vessel number, structure, function, direction of flow and relationship to the location of fetal trachea can also be examined. In addition to allowing better comparison between vessels, these views are easier to achieve and this reduces the time need to visualise the outflow tracts when compared to traditional long-axis view.<sup>27</sup> Transverse views of the fetal heart are sufficient to identify all the normal features and to exclude cardiac malformations.

#### **Recognising abnormalities in the Three-Vessel View**

Most lesions involving the ventricular outflow tracts and/or Great Arteries are associated with an abnormal Three-Vessel View.<sup>29-31</sup> Abnormalities that can be visualised include abnormal vessel size, number, alignment, abnormal flow or spatial relationship of the aortic arch. An abnormal Three-Vessel View raises the suspicion of abnormality and requires a detailed fetal echocardiogram, both 2D and color Doppler to make a final diagnosis. It also provides an important clue to the diagnosis of minor anomalies considered as markers of other CHD.

#### Abnormal vessel size

In the normal Three-Vessel View, the pulmonary trunk has a slightly larger diameter than the aorta, which has a larger diameter than the superior vena cava. Each of these vessels may by dilated or constricted, which can be recognised by direct



Figure 5: Arch View, Coarctation of aorta.



**Figure 6:** Interrupted aortic arch, note the disproportion between pulmonary artery (PA) and aorta (Ao).

comparison during grey scale imaging and with the help of color Doppler. Obstruction of either the aortic or pulmonary valve will prevent forward flow of blood and the respective Great Arteries will have an abnormal ratio of diameters.<sup>32</sup>

#### Small Aorta

A small ascending aorta or a small/absent transverse aortic arch is seen in fetuses with coarctation of aorta (CoA), Interruption of aortic arch (IAA), aortic stenosis and aortic atresia.

CoA is the most common duct dependent cardiac abnormality and is one of the most difficult cardiac abnormalities to diagnose in utero. It accounts for approximately 7% of all live births with congenital heart disease. An estimated 60% to 80% of newborns with isolated CoA are discharged home without diagnosis and they face circulatory collapse and death as the duct closes.<sup>33–36</sup> Current screening methods detect < 30% cases with a high falsepositive rate (50–60%) even in the hands of fetal cardiologists.<sup>37,38</sup>



Figure 7: Small pulmonary artery.

The aortic isthmus region, just before where the arterial duct enters the descending aorta, is the narrowest part in CoA.<sup>39</sup> The three vessel and trachea view allows direct comparison of the aortic and ductal arches in size and easy assessment of the fetal aortic isthmus.<sup>40–43</sup> Although ventricular disproportion (with a smaller left than right ventricle) on a 4 chamber view raises the suspicion of the diagnosis, the sensitivity of ventricular disproportion is only moderate and the specificity is poor, especially after 34 weeks. Four-chamber disproportion may be subtle, and disproportion may be visible only at the arch views (Figures 4, 5). Therefore, screening using the Three-Vessel View is important to avoid false-negative results.

Interruption of the aortic arch (IAA) is a rare anomaly in the fetus and neonate and occurs in 3/100,000 live births and is rapidly fatal if left uncorrected.<sup>43</sup> IAA is classified into three morphologic types, but type B, interruption is between the left carotid and left subclavian arteries, is the most important. Ventricular septal defects are commonly seen in association with IAA but prenatal detection of IAA by echocardiography has been challenging.

The Three-Vessel View is the best view to diagnose IAA. A large discrepancy in the size of the aorta and pulmonary artery is a pathognomonic sign of IAA type B (Figure 6).<sup>44,45</sup> Sometimes it is difficult to differentiate this from severe CoA but IAA almost always have a large ventricular septal defect, there is no ventricular disproportion and the transverse aortic arch can not be visualised in a Three-Vessel View, in contrast to CoA where disproportion and a small transverse aortic arch are usually recognisable.

In aortic atresia there is complete obstruction of the blood flow from the left ventricle and aorta is very hypoplastic and thread like. This can be an isolated condition but usually found in association with mitral atresia. In Three-Vessel View the aorta can be difficult to visualise on 2 Dimensional imaging however color Doppler imaging demonstrates reverse flow through the arterial duct.



Figure 8: Simple transposition of Great Arteries.

#### Small pulmonary artery

A small pulmonary artery means pulmonary artery is receiving less blood flow and this could be because of pulmonary stenosis or atresia or alternatively less blood flow because of obstruction further downstream ie tricuspid valve. This is not a specific finding and can be found in Tetralogy of Fallot (TOF) with pulmonary stenosis, in some forms of double-outlet right ventricle with pulmonary stenosis, or in Ebstein's anomaly or Tricuspid atresia.

Tetralogy of Fallot (TOF) occurs in approximately 3/10,000 live births and is frequently associated with chromosomal anomaly and extra cardiac malformations.<sup>46</sup> Fetal diagnosis is based on presence of large ventricular septal defect, aortic overriding and small pulmonary artery and a relatively large aorta.<sup>47</sup> Usually 4-chamber view is normal, however there can be a left shift of the cardiac apex.

The clinical symptoms and requirement of early intervention depends upon the degree of right outflow tract obstruction. Most patients with typical TOF undergo corrective surgery at around 3 to 6 months of age with good functional result. Those with severely obstructed pulmonary blood flow, fetal diagnosis allows better planning, facilitates early prostaglandin therapy to maintain ductal patency, therefore avoiding life-threatening cyanosis in the early newborn period.

Current routine screening has a detection rate of 15–40%.<sup>23–</sup> <sup>25</sup> The diagnosis of TOF can't be made unless outflow tracts are carefully evaluated.

The aortic overriding and ventricular septal defect can be missed on initial screening however Three-Vessel View is always abnormal with pulmonary artery being smaller in size as compare to aorta and recognition of this leads to more careful examination of the septum (Figure 7). The size of the main pulmonary artery and its ratio to the ascending aorta reflects the severity of outflow tract obstruction, being significantly smaller in those who will present as a duct-dependent pulmonary circulation.



Figure 9: Reverse flow in the aortic arch.



Figure 11: Persistent left superior vena cava.

#### Superior vena cava

The superior vena cava can be enlarged in conditions like supra cardiac total anomalous pulmonary venous connection, aneurysm of the vein of Galen, when there is increased venous return from the brain or because of an increased blood flow in the vessel when azygous vein is dilated in conditions with interrupted inferior vena cava.<sup>31,48</sup>

#### Abnormal vessel number – Two vessels in Three-Vessel View

The absence of one great artery can be demonstrated in the Three-Vessel View, leading to suspicion of several defects, such as truncus arteriosus, TGA, aortic atresia and TOF with pulmonary atresia.

Simple transposition of the Great Arteries is diagnosed by finding the aorta arising from the right ventricle and pulmonary artery arising from the left ventricle. This occurs in 1/3500–5000 live births and accounts for 4–7% of all congenital heart diseases.<sup>49</sup> Simple transposition can be successfully treated



Figure 10: Reverse flow in the pulmonary artery.



Figure 12: Isolated left superior vena with absent right superior vena cava.

by arterial switch operation in the first week of life, with good surgical results and low mortality.

About 10–20% of cases of transpositions require urgent treatment after birth because they have an inadequate size of patent atrial communication, which is essential in this condition for "mixing" and survival.<sup>49</sup>

Despite including outflow tracts in routine screening the detection rate of TGA remains low (15-52%).<sup>23-25</sup> A normal Three-Vessel View is impossible to obtain in transposition as the pulmonary trunk lies underneath the aortic arch and only two vessels are seen in three-vessel view (Figure 8).<sup>50</sup>

Sometime a very small aortic arch in aortic atresia and a small pulmonary artery in pulmonary atresia may not be visible on two dimensional ultrasound giving the appearance of two vessels in Three-Vessel View and color Doppler helps to identify the hypo plastic arch showing retrograde flow from the isthmus (Figure 9) and pulmonary atresia with reverse flow through the ductus arteriosus (Figure 10).



Figure 13: Right sided aortic arch.

# aortic arch.

*Four vessels in Three-Vessel View* Four vessels instead of three are seen in Three-Vessel View in cases of a persistent left superior vena cava (PLSVC) (Figure 11). Isolated PLSVC is a normal variant of vascular anatomy and found in 1:300 routine scans and it has no postnatal hemodynamic consequences.<sup>51–53</sup> The persistent LSVC usually drains to the coronary sinus, which becomes enlarged and may be associated with disproportion at the four-chamber view. In almost all conditions where there is bilateral superior vena cava there is an absence of the bridging vein, namely the innominate or left brachiocephalic vein.<sup>54,55</sup> The presence of left superior vena cava with absence of the right superior cava is more challenging to diagnose as in this condition, three vessels are identified but the SVC on the right is absent and this is found on the left of the pulmonary artery (Figure 12).

# Abnormal vessel arrangement

A normal Three-Vessel View demonstrates that the left aortic arch crosses from right to left in front of the spine and forms a V shape. When the transverse aortic and ductal arches do not form a typical V-shape pointing to the left with the superior vena cava to their right, the Three-Vessel View is considered abnormal.

In anatomical terms, a right aortic arch (RAA) is one where the arch crosses over the right bronchus, instead of the left as normal. The incidence of RAA in the general population is unknown, but a rate of 1/1000 in low-risk pregnancies has been suggested.<sup>56</sup> It is a normal variant of vascular anatomy. A RAA is found in about 25% of all conotruncal anomalies and its detection should lead referral for a comprehensive fetal echocardiogram.<sup>56-61</sup>

Usually an isolated RAA has no hemodynamic consequences unless it forms a vascular ring in association with aberrant vessels or double aortic arch.

In fetal life, the diagnosis of the RAA is made from the relationship of the transverse portion of the aortic arch to the trachea, with the arch crossing from right to left in front of the trachea in normal cases, or remaining on the right of it in a RAA.



Figure 14: The right aorticRight aortic arch and Right duct.

An aortic arch descending on the right side of trachea is easy to appreciate in the Three-Vessel View (Figure 13). Demonstration of the position of the aortic arch is difficult in long-axis views. A RAA may also be found in combination with a left sided or right-sided arterial duct. RAA in combination with left arterial duct with the trachea lying between them produces a 'U-sign'. The RAA in combination with right arterial duct can be challenging to diagnose as it still forms a V shape however this is on the right side of the trachea (Figure 14).

# Abnormal flow

Outflow tract views are more sensitive than the 4CH view in detecting duct dependent cardiac anomalies, which are the ones that benefit the most from prenatal detection. Both Great Arteries should have forward flow on color Doppler imaging. Reverse flow in either the aortic arch or the duct indicates postnatal duct dependency. Reverse flow in the aortic arch is found in cases of aortic atresia, severe aortic stenosis and severe coarctation of aorta (Figure 9). Reverse flow in the pulmonary artery is found in cases of pulmonary atresia and severe pulmonary stenosis (Figure 10).<sup>62</sup> Aliased flow in the aorta or pulmonary artery may reflect an increased pressure across a stenotic semilunar aortic or pulmonary valve.

# Conclusion

As the majority of pregnant women (> 70%) have their routine anomaly scans in non-tertiary settings, the screening examination should be simple and sensitive. At least 20–25% of neonates are discharged home without a diagnosis and some die before their congenital heart defect is recognised. One important consequence of prenatal diagnosis is the avoidance of hemodynamic decompensation in duct-dependent lesions, such as critical left heart obstructive lesions, transposition of the Great Arteries, and pulmonary atresia.<sup>21</sup> Routine fetal heart screening in the mid second trimester should include imaging the heart in a transverse section from the level of the diaphragm to the inlet of the thorax –five essential views and include the transverse section of abdomen, four-chamber view, the outflow

tracts, and the three-vessel and the transverse arch view. The Three-Vessel View is easy to acquire and interpret, and adds more to the screening than the long axis views of the aortic and ductal arches. Long axis views of the archs are not required for routine screening. However, once an abnormality is detected, it should be confirmed in long axis views.

#### Key points:

- CHD is the most common structural malformation.
- 85% of CHD occurs in low risk group.
- Detection rate can be increased only by routine screening of low risk population.
- Five essential views in transverse section are required for routine screening of CHD.
- Long axis views are not required for routine screening.

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