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Associated clinical guidelines/protocols:
- Cardiac ICU guidelines

Information for Year 1 ITU Training (basic):

Year 1 ITU curriculum
- Understand the ITU post-op management and common complications of Right ventricular outflow tract obstruction
  - Tetralogy of Fallot
  - Critical Pulmonary Stenosis
- Principles of Echocardiography and Cardiac catheterization relevant to the above lesions.

Curriculum notes for year 1:

Understand the ITU post-op management and common complications of:
Right ventricular outflow tract obstruction

Cyanotic congenital heart disease with decreased pulmonary blood flow in children

1. Tetralogy of Fallot
   1.1 Anatomy and Physiology
   1.2 Palliation with BT shunt
   1.3 Associated Anomalies
   1.4 Total Surgical Repair
   1.5 Post op issues
   1.6 Restrictive RV physiology
2. Critical Pulmonary Stenosis
3. Tetralogy of Fallot with Pulmonary Atresia
4. Pulmonary Atresia with intact ventricular septum (IVS)
5. Tricuspid Atresia
6. Ebstein’s anomaly

The first two lesions are covered in Year 1 and the rest in Year 2 curriculum.
TETRALOGY OF FALLOT

This is a condition where the subpulmonary conus fails to expand normally. The anatomical hallmark is the subpulmonary infundibular narrowing due to the anterior and cephalad deviation of the outlet septum relative to the septomarginal trabeculation. This single feature accounts for the VSD and the rightward position of the aortic root.

PRE-OPERATIVE ANATOMICAL AND CLINICAL CONSIDERATIONS

The classic tetrad (Figure 1) consists of
- Overriding aorta
- Right ventricular hypertrophy
- Subpulmonary stenosis
- Malalignment ventricular septal defect

Figure 1a: Tetralogy of Fallot – Anatomy – Picture taken from Prof Anderson’s Cardiac Core Curriculum Training Slides

Figure 1b: Tetralogy of Fallot

The anatomy of Tetralogy of Fallot. The key issues are schematically shown. As noted by Anderson and Weinberg, components of the supraventricular crest have “sprung apart.” The cardinal feature is the anterocephalad deviation of the outlet septum. The secondary necessary feature is hypertrophy of the septoparietal trabeculations in order for subpulmonic obstruction to be present. The other classic features include RV hypertrophy, an interventricular communication (VSD), and an overriding Aorta (shown through the VSD).

Figure 1b. Image from Bashore et al. Adult Congenital Heart Disease. Right Ventricular Outflow Tract Lesions. Circulation. 2007;115:1933-1947
Physiology

The anatomy of the TOF determines the physiology.
- The right ventricular pressure is the same as the left ventricular and aortic pressures as a result of the large unrestrictive VSD.
- The pulmonary artery (PA) pressure and flow are inversely proportional to the degree of subpulmonary obstruction.
- Greater stenosis (or hypoplasia) causes lower pulmonary artery pressure, less pulmonary blood flow and increasing right ventricle through VSD-to-aorta flow.
- The degree of cyanosis correlates precisely with the amount of obstruction, which being muscular can be highly dynamic and variable.
- The basic anatomy can range from one end of the spectrum with mild subpulmonary stenosis to severe stenosis and atresia.

REPAIR

Evolving approach from 2 stage – initial palliative systemic-pulmonary shunt and then complete repair to a higher risk primary repair in children fewer than 3 months of age.

Modified Blalock Taussig shunt – involves insertion of a polytetrafluoroethylene tube graft on the side opposite to the descending aorta from the proximal subclavian artery or distal innominate artery, side to side to the ipsilateral pulmonary artery. The size of the graft (3.0, 3.5, 4.0, 5.0) depends on patient size and calibre of pulmonary artery.

Indications in TOF

1. Anomalous major coronary artery
2. Long and narrow ventricular outflow
3. Association with complete AVSD
4. Multiple VSD’s
5. Severely hypoplastic pulmonary arteries

Figure 2: Systemic to pulmonary artery shunt

Figure 2. Image from Bashore et al. Adult Congenital Heart Disease. Right Ventricular Outflow Tract Lesions. Circulation. 2007;115:1933-1947
Advantages
- Highly reliable
- Does not distort the pulmonary arteries
- Can be matched in size to the patient and the pulmonary artery
- Has not been reported to cause pulmonary vascular obstructive disease
- Is easy to take down
- Placement does not require CPB and can be done either thro’ a median sternotomy or a lateral thoracotomy.

OTHER ASSOCIATED ABNORMALITIES in TOF
To be considered before complete repair
- Coronary artery origin and any anomalies
- Aortopulmonary collaterals
- Length of RVOT obstruction
- Size of Pulmonary Arteries
- VSD extension and other VSD’s
- Associated defects like mitral stenosis, PDA, ASD or AV canal defect
- Previous shunts

Coronary anomalies
Approximately 5% with TOF have the left coronary arising from the RCA and then coursing across the RVOT to distribute the septum and the left ventricle. An incision into the RVOT may damage this vital coronary artery. In addition, there are many variations of the coronary artery origin and course e.g. dual LAD supply, which can be potentially harmed during surgery.

Aortopulmonary collaterals - Large collateral vessels connecting the aortic tributaries or the descending aorta with the branch pulmonary arteries may be present, which can produce a large, left to right shunt. These may be incorporated into the repair of the pulmonary arteries or they may be closed surgically or by interventional catheter.

Other abnormalities
1. Additional VSD – A second inlet muscular VSD
2. Associated ASD – called as ‘Pentalogy of Fallot’
3. Right-sided aortic arch - present in 25%; 90% have mirror image branching and 10% may have aberrant LSCA.
4. Persistent left SVC
5. Left sided lesions – rarely mitral stenosis may be present in TOF.

Other associated lesions of the pulmonary circulation
1. Pulmonary valve stenosis – though the classic form of RVOTO is infundibular stenosis – 75% have both infundibular and pulmonary valve stenosis.
2. Pulmonary artery and branch PAs - Pulmonary artery and supravalvar stenosis and branch PA hypoplasia/ataresia are frequently associated and are a consequence of decreased pulmonary blood flow during development. Lack of origin of one pulmonary artery (typically the left) from the pulmonary trunk is not infrequent. The non-connected PA (usually the left branch pulmonary artery) can at times originate from the PDA.
3. TOF with diminutive pulmonary arteries – these may be treated with outflow tract patch without VSD closure. It is hoped that the increased forward flow into the pulmonary artery system induced by the relief of obstruction will encourage pulmonary artery growth so that the VSD can be closed later.
4. TOF with absent pulmonary valve syndrome – discussed separately later in year 2 curriculum.
5. TOF with pulmonary atresia – discussed separately later in year 2 curriculum.
6. AVSD and TOF – seen exclusively in children with Trisomy 21. The combination of a large VSD (both inlet septal defect and malalignment) and the valvar abnormalities
(pulmonary and atrioventricular) complicate reparative surgery and increase the risk and likelihood of postoperative morbidity.

7. **Aortic root dilatation** is a known clinical feature of unrepaired TOF. In TOF patients, aortic dilatation occurs predominantly in the aortic root and proximal ascending aorta, with relative sparing of the aortic arch, and descending thoracic and abdominal aorta. The underlying pathophysiology was initially attributed to altered hemodynamics resulting from longstanding volume overloading and stretching of the aortic root from increased right to left shunting. Recent review suggested cellular expression of an unrecognized gene associated with conotruncal defects as a cause of the dilatation. Findings of intrinsic histological abnormalities in the aortic root and ascending aorta of tetralogy of Fallot patients suggest that intrinsic abnormalities may also play an important causative role.

**COMPLETE REPAIR OF TETRALOGY OF FALLOT**

**Advantages**

1. Prevents development of right ventricular hypertrophy.
2. Early repair provides adequate and homogenous pulmonary blood flow and allows normal growth and development of pulmonary vessels (Kolcz et al 2005, Van Dongen et al 2003)

**PRINCIPLES IN COMPLETE SURGICAL REPAIR** (see Figure 3)

- Closure of VSD
- Remove RVOT obstruction
- Avoid conduction system (Figure 4)
- Look for any abnormal coronary branch crossing RVOT

**Figure 3. Classic repair in tetralogy of Fallot.** An outflow patch of pericardium is used to relieve subpulmonic obstruction, and the VSD is closed with a pericardial or Dacron patch. If valvular PS is present or the main PA is hypoplastic, the outflow tract is extended into the main PA.

**Removing RVOT obstruction**

3 methods
- Remove subvalvar obstruction with or without pulmonary valvotomy – leaves residual RVOTO
• The first option plus transannular patch or annular patch – creates PR
• Insertion of orthotopic valve or valve containing conduit – non growing element left in the heart

Surgical approach

• Combined transatrial and transpulmonary approach is preferred to the traditional transventricular approach.
• Subvalvar infundibular incision is used when the pulmonary annulus is adequate.
• Transannular incision is used when the pulmonary annulus is inadequate.
• Pulmonary valvotomy
• Resection of hypertrophied septal and parietal bands of the infundibulum to relieve the infundibular stenosis (major site of RVOTO).
• RVOT patch – preservation of PV annulus
• RVOT patch with resection of infundibular muscle (augments restrictive pulmonary blood flow)
• Patch closure of VSD
• Transannular patch to augment the RVOT at the levels of the infundibulum, valve annulus and main pulmonary artery. Disrupts integrity of pulmonary valve annulus and results in free PR. It is used when the PV annulus is restrictive.

POSTOPERATIVE ISSUES

Residual VSD
May be poorly tolerated in the presence of co-existing pulmonary regurgitation, non-compliance of the ventricles, a left ventricle not previously volume loaded or a combination of all these factors. Patients with haemodynamically important residual VSD have inappropriately elevated heart rate and high left atrial pressure (LAP). The LAP may be significantly higher than the right atrial pressure. The presence of residual VSD needs prompt attention and repair if significant.

Residual outflow tract obstruction
Though mild residual RVOTO is well tolerated in the immediate postoperative period, it can lead to ventricular arrhythmias over time and the need for re-operation.
LCOS after TOF repair
Causes - May be due to a) residual lesions b) arrhythmia c) restrictive RV physiology d) Post CPB nadir in cardiac index.

RV dysfunction
Ventricular dysfunction may be related to the combination of diastolic dysfunction (restrictive RV physiology), the right ventricular incision and pulmonary regurgitation. It is important to rule out residual VSD and or RVOTO.

Rhythm abnormalities
- JET (diagnosis and management described in module on Arrhythmias)
- Right Bundle branch block - Diagnosed by increased duration of QRS complex (>0.12 secs), rSR' pattern in QRS complex in V1 and/or V2, (large terminal R' at the end of the QRS complex as the RV is depolarizing slowly without the special conduction of the right bundle branch via myocardial routes outside of the His-Purkinje system) and slurred S waves in I, II, V5 and/or V6 (indicates late right ventricular depolarization in a direction away from the orientation of the lead).

It is seen in nearly 50% of patients undergoing TOF repair. The course of the bundle branches varies with the position of the VSD, which has a direct bearing on the incidence of postoperative bundle branch block. It is unclear whether the postoperative RBBB is caused by the ventriculotomy, damage to moderator band, VSD repair itself or resection of infundibular muscle. There are no adverse prognostic indicators seen in patients with RBBB after TOF repair versus those without RBBB.
- Bifascicular block
- Complete heart block

Restrictive RV Physiology

Characteristics - Restrictive RV physiology is characterised by diastolic dysfunction where end-diastolic pressure is higher at any given end-diastolic volume as a result of the loss of ventricular compliance. It is generally seen in patients with hypertrophied ventricle undergoing surgical repair and in neonates when myocardial oedema may significantly restrict diastolic dysfunction.

Causes - The origin of the acute RV restriction after TOF repair is not known. The superimposition of the effects of cardiopulmonary bypass, ventriculotomy, myocardial oedema and the placement of non-functional patches on the ventricular septum and in the right ventricular outflow tract on the background of right ventricular hypertrophy - all might be expected to influence the diastolic performance of the right ventricle. Intra-operatively, the RV may not be not be adequately protected, because its anterior position makes satisfactory hypothermia difficult and hypertrophy complicates the homogeneous delivery of cardioplegia. Endomyocardial fibrosis has also been demonstrated in older patients with Tetralogy of Fallot.

Diagnosis – Restrictive RV physiology can be diagnosed on echocardiography. The hallmark of isolated severe right ventricular restriction is antegrade diastolic pulmonary blood flow coincident with atrial systole associated with increase in RV pressure. The right ventricle becomes unfillable during atrial systole and acts as a passive conduit. Thus, some or all of the tricuspid atral systolic flow, demonstrable by Doppler, results in antegrade pulmonary blood flow, not right ventricular filling. The simultaneous retrograde flow in the superior vena cava and rapid deceleration of the early rapid filling velocity seen in many patients are consistent with a restrictive right ventricle. It is important to recognize that under these circumstances, traditional indices of restrictive physiology may not be measurable (because of summation flow related to tachycardia) or valid such as atrial "filling" fraction because not all tricuspid flow equates to filling. Right ventricular systolic function is qualitatively normal in these patients with almost complete obliteration of the right ventricular cavity at end systole in most of the patients. Doppler indexes of left ventricular function are normal in patients with restrictive right ventricular physiology despite paradoxical septal motion.
**Restrictive RV and positive pressure ventilation** - Cullen et al. demonstrated that this pulmonary arterial antegrade flow coincident with atrial systole was augmented during the expiratory phase of positive pressure ventilation and abolished or greatly diminished during the inspiratory phase \((P<0.001)\). An increase in the duration of pulmonary regurgitation occurred during the inspiratory phase of positive pressure ventilation in these patients \((P<0.01)\).

**Clinical features** - Patients with restrictive right ventricles have clinical features of a low cardiac output, require higher filling pressures, and although biventricular systolic performance is well preserved, they often require prolonged inotropic and volume support. This leads to a significantly greater incidence of effusions and a longer stay in intensive care. Although reflecting adverse haemodynamics, the antegrade diastolic pulmonary arterial flow seen in these patients is important as it contributes to forward flow and shortens the duration of pulmonary regurgitation, making an important contribution to cardiac output. Conversely, anything that diminishes this effect will be detrimental. Thus, maintenance of sinus rhythm is important in patients with restrictive physiology, and the possible beneficial effects of changes in atrioventricular delay by atrioventricular pacing warrant further study. Furthermore, transient loss of these beneficial effects is seen during the inspiratory phase of positive pressure ventilation. It is therefore recommended that intermittent positive pressure ventilation with a short inspiratory time and lowest possible mean airway pressure be used in these patients.

**Treatment of RV restrictive physiology involves reduction of RV afterload**

- Ventilatory manoeuvres – early extubation, spontaneous breathing, ventilation at FRC, and extrathoracic negative pressure ventilation.
  - Shekerdemian et al. showed that a brief period of negative pressure ventilation in the early postoperative period after complete correction of TOF increased pulmonary blood flow by 39%, and the improvement further continued if the study period was extended, with a total increase of 67% after 45 min. Patients with restrictive physiology had a somewhat delayed response to NPV, but the ultimate increase during an extended period of NPV was greater in restrictive patients (84%) than in non-restrictive patients (50%).
- Avoid tachycardia, maintain AV synchrony
- Reduce catecholamines
- Use vasodilators - to alter systolic wall tension and decrease the impedance to ventricular ejection. Milrinone – inodilator with lusiotropic and vasodilator properties is especially useful.
- Volume replacement – to maintain adequate filling pressures

**Aortic Root dilatation**

Among the postrepaired TOF patients, there is a subgroup of patients with progressive aortic root dilatation. In these patients, the aortic root and ascending aorta may dilate insidiously, leading to aortic regurgitation requiring aortic valve replacement. In addition, the dilated aortic root and ascending aorta are also at risk of dissection and rupture.

**CRITICAL PULMONARY STENOSIS**

In common usage, pulmonary stenosis (PS) is the presence of a well-formed right ventricle and tricuspid valve, whereas pulmonary atresia (with IVS) implies hypoplasia of the tricuspid valve and right ventricle. The natural history and therapeutic options of critical PS are different from pulmonary atresia. In neonates with critical PS, the right ventricle fills with the normal volume of blood, most or all of which must return to the right atrium via tricuspid regurgitation because of the severe obstruction at the pulmonary valve. This reduces direct flow into the lungs and forces the large atrial right to left shunt. Pulmonary blood flow is ductal dependant. When the interatrial communication is restrictive, these cyanotic infants may also demonstrate dramatic right atrial enlargement and hepatic venous congestion. The left heart may appear small because of under filling and in comparison to the hypertrophied
right side. Again, in contrast to children with TOF, those with critical PS always have well formed pulmonary artery systems. Most children with critical PS have interventional catheter repair. When any opening in the pulmonary valve is present, a wire is advanced through this orifice and balloon dilatation is performed using first a small balloon (5-6 mm) and then larger sizes 8-12 mm. When the valve is atretic, a stiff wire is used under echocardiographic guidance in the catheterisation lab to perforate the membrane after which the balloon dilatation technique is used. Because these children generally have adequate sized tricuspid valves and right ventricles, a balloon atrial septostomy is not performed because it can be potentially harmful. The hypertrophied right ventricles of children with critical PS are usually poorly compliant and need good preload, anything that reduces right ventricular filling such as decompression through a large ASD, reduces both pulmonary blood flow and the stimulus to normal right ventricular growth. The outlook for children with critical PS is excellent when normal forward flow is established and the right heart can function normally.

Websites.
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References

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Curriculum notes for year 2:

TETRALOGY OF FALLOT WITH PULMONARY ATRESIA

TOF with pulmonary atresia is at times referred to as pulmonary atresia with VSD. However, the outlet septum in this lesion is deviated in a manner reminiscent to that of classic tetralogy of Fallot, in fact so severely so as to cause complete right ventricular outflow tract obstruction. Hence, the central feature in the tetralogy of Fallot as related to antero-cephalad deviation of the outlet septum and the abnormal relationships to the ventriculoinfundibular fold and septomarginal trabeculation are also present in this lesion. The proper terminology for this lesion should therefore be that of tetralogy of Fallot with pulmonary atresia. TOF/PA is an extremely heterogeneous lesion not because of the actual cardiac anatomy but because of the variability of pulmonary blood supply.

Figure 5: TOF with PA

Morphology & Embryology

Early during foetal development, the vascular plexus within the lung buds connects with systemic segmental arteries originating from the dorsal aorta. By the 40th day of gestation, the vascular plexus has differentiated into pulmonary segmental arteries, supplying the terminal bronchopulmonary units. For a short time, the pulmonary parenchyma receives a dual blood supply (from the right ventricle and the pulmonary arteries that originate from the sixth branchial arches and from the previously described systemic segmental arteries). However,
by the 50th day of gestation, the systemic arterial supply normally involutes, and during subsequent normal fetal development, flow to the developing lungs is delivered exclusively by the pulmonary arteries.

In the more complex forms of TOF with PA, this normal development is affected, whereby some bronchopulmonary segments are supplied by true pulmonary arteries, and others by aorto-pulmonary collaterals. It is important to note that before entering the lung parenchyma, these systemic collaterals retain their histologic characteristics of muscular arteries; whereas after penetrating the pulmonary parenchyma, the medial muscular layer gradually changes into an elastic lamina, structurally resembling true pulmonary arteries. Unobstructed flow through aorto-pulmonary collaterals can lead to pulmonary vascular obstructive disease, while stenosis within aorto-pulmonary collaterals protects against the development of pulmonary vascular obstructive disease.

The pulmonary atresia may be found at the level of the subpulmonary infundibulum, in which case it is often an acquired lesion, or more commonly, at the level of the muscular septum or pulmonary annulus, in which case the cause is likely to be congenital in origin. The ventricular septal defect is usually perimembranous, but can also have a muscular postero-inferior rim. Both the subpulmonary infundibulum and the outlet septum may be completely missing, in which case both the ventricular septal defect and outflow tract is reminiscent of that of truncus arteriosus.

When tetralogy is accompanied by pulmonary atresia, the determinant of clinical presentation and prognosis is the source of the pulmonary blood flow, which under these circumstances can be derived from either a persistent arterial duct or from aorto-pulmonary collaterals.

**Characteristics of pulmonary blood flow (Figure 6)**

- When pulmonary blood flow is derived from a persistent arterial duct, then the branch pulmonary arteries are usually confluent and the duct is left-sided, irrespective of which side the aortic arch is located.

- When the branch pulmonary arteries are nonconfluent, which is rare in the presence of a patent arterial duct, each branch pulmonary artery may be supplied by a one of a bilateral pair of arterial ducts.

- In the presence of duct-dependent pulmonary blood flow, aorto-pulmonary collaterals are usually clinically insignificant irrespective of their presence or number.

- When pulmonary blood flow is derived from aorto-pulmonary collaterals, the anatomy is much more complex.
  - Aorto-pulmonary collaterals most frequently arise from the descending aorta and vary in number from two to six. They may also arise from the brachiocephalic arteries, or rarely, from the coronary arteries.
  - Almost always, aorto-pulmonary collaterals coexist with intrapericardial pulmonary arteries, in which case they anastomose within the parenchyma of the lungs.

- It is important in planning a course of ultimate unifocalization to determine the source of arterial blood supply for each segment of lung, namely whether it is derived from an intrapericardial pulmonary artery or whether it is derived from an aorto-pulmonary collateral.

- In some cases of nonconfluent pulmonary arteries, one lung may be supplied by aorto-pulmonary collaterals, while the other lung is supplied by a single branch pulmonary artery derived from either the arterial duct, or directly from a systemic blood source.

**There are 3 main subgroups**

1. Confluent through i.e. mediastinal pulmonary arteries, normal to slightly small in calibre supplied by the ductus.
2. Absent or extremely diminutive <2mm two pulmonary arteries with multiple aorto-pulmonary collateral arteries (MAPCA’s)

3. Small mediastinal pulmonary arteries and MAPCA’s with multiple segments of lung receiving dual supply.

Preoperative Diagnosis
The diagnostic challenge in TOF with PA is to identify preoperatively the presence, size, and continuity of native pulmonary arteries and then to detail the origin, the course, and the distribution of all aorto-pulmonary collaterals. It is virtually always necessary to make selective injections into all direct and indirect aorto-pulmonary collaterals to obtain a complete and detailed map of the entire pulmonary blood supply.

Accurate measurement of the size of a pulmonary artery preoperatively presents a number of problems. First, with diminished pulmonary blood flow, the maximal capacity or compliance of the non-distended pulmonary arteries cannot be accurately assessed. Consequently, the potential postoperative size of a pulmonary artery carrying a normal volume of blood is difficult to predict. Nevertheless, several methods to quantify pulmonary artery size and its effect on the post-repair outcome have been used.

Formulae to help quantify pulmonary artery size

McGoon ratio - It is based on the diameter of the right and left pulmonary arteries, normalizing these by relating them to the diameter of the descending thoracic aorta at the level of the diaphragm. Right and left pulmonary arteries are considered to be nonrestrictive.

Figure 6: Systemic sources of pulmonary blood supply in PA with VSD.
Figure from textbook of Paediatric Cardiology. Eds Anderson, Maccartney, Shinebourne, Tynan. Churchill Livingstone
when the combined diameter is about 2 or greater, while a combined diameter of less than 0.8 is supposed to indicate severely restrictive central pulmonary arteries. One drawback of the McGoon ratio is that the descending aorta at the diaphragm tends to be narrower in patients with Tetralogy of Fallot than in normal individuals, making the McGoon ratio falsely more favourable.

**Nakata Index** - Nakata and colleagues measured the diameter of the right and left pulmonary arteries immediately proximal to their first branching. Magnification errors are corrected either by using previously determined values from the catheterization laboratory or by relating vessel size to the known size of an appropriate catheter. Pulmonary artery size is reported as the sum of the cross-sectional areas of the right and left pulmonary arteries, indexed to body surface area. The normal cross-sectional index is $330 + 30 \text{ mm}^2/\text{m}^2$, and is considered diminutive when the Nakata index is less than $150 \text{ mm}^2/\text{m}^2$.

**Z scores** - Normalization of the dimensions of cardiac structures to the size of the body, using so-called Z scores, is being increasingly used in the management of congenital heart disease. Regression equations are derived relating cardiac dimensions (measured using cross-sectional echocardiography in normal infants and children) to the size of the body. The expression of size with the highest correlation to cardiac dimensions was body surface area. Nomograms were then developed from which the Z score of a cardiac structure could be estimated from knowledge of the body surface area and the echocardiographically derived measurement.

Using statistical techniques, Blackstone and colleagues predicted the postoperative $P_{RV/LV}$ based on the dimensions of the right ventricular outflow tract and the size of the central branch pulmonary arteries (and not the peripheral pulmonary artery branches). If the pulmonary valve annulus is hypoplastic, it tends to remain so throughout life. Therefore, Blackstone and Kirklin expressed the annular size relative to the child's size as a Z value, which represents the number of standard deviations that the patient's pulmonary valve annulus deviates from a mean normal value for age and size.

All of these assessments including the angiographic ones are limited in that the size of the pulmonary arteries may enlarge significantly after establishing right ventricular to pulmonary arterial continuity, given the increased volume and distending pressure. On the other hand, there clearly is a subset of patients in whom the central branch pulmonary arteries are too diminutive in size, generally less than 3 mm in diameter, that they cannot carry right ventricular output, thereby contraindicating ventricular septal defect closure.

Most patients with tetralogy of Fallot with pulmonary atresia and a duct-dependent pulmonary circulation have sufficiently large pulmonary arteries, (generally with a Nakata index greater than $150 \text{ mm}^2/\text{m}^2$) that they can be successfully repaired at a low operative risk with good late haemodynamic and electrophysiological results.

A therapeutic challenge is presented in the subset of patients with diminutive pulmonary arteries, generally with a Nakata index less than $100 \text{ mm}^2/\text{m}^2$, and large aorto-pulmonary collaterals that supply a variable number of bronchopulmonary segments. The ultimate therapeutic goal in this subset of patients is to establish right ventricular-dependent pulmonary circulation, which would ideally include all 20 bronchopulmonary segments. Haemodynamically, the aim is to achieve a postoperative $P_{RV/LV}$ ratio of less than 0.6 with no residual left-to-right shunt at any level. Until relatively recently, this ideal result has been achieved only in isolated cases, primarily due to limitations of surgical technique in dealing with these complex anatomic features. Recently it has become evident that many of these complex lesions, including the presence of dual-supply segments and stenoses can be managed with by invasive interventional techniques, and by aggressively treating these lesions during early infancy.

**Management**

**Staged intervention with a goal to reconstruct an adequate pulmonary artery tree to accommodate the entire cardiac output at acceptably low pressures.**

- If confluent i.e. mediastinal pulmonary arteries, normal to slightly small in calibre supplied by the ductus - manage as TOF until either a palliative shunt is performed or alternatively complete correction can be performed.
Left PA artery stenosis at the ductal insertion site is common in this type. For complete correction, a right ventricular pulmonary artery conduit is typically necessary. If complete neonatal repair is undertaken, the distal end of the conduit can be incorporated in such a way to plasty the left pulmonary artery origin. The VSD can be closed with a ventriculotomy that is used to attach the proximal end of the conduit.

- If absent or extremely diminutive (<2mm) pulmonary arteries with multiple aorto-pulmonary collateral arteries (MAPCA’s) of if small mediastinal pulmonary arteries and MAPCA’s with multiple segments of lung receiving dual supply. The approach is more individualised depending on the particular anatomy of the coronary arterial tree. It generally involves establishing forward flow into the true pulmonary arteries in the mediastinum followed by angiography to determine the segments of lung that are supplied by a) the true pulmonary arteries b) MAPCA’s alone and c) both. Segments supplied by MAPCA’s may need to be unifocalised into the true pulmonary artery confluence. There may be segments with proximal stenosis but are supplied by the true pulmonary arteries, which may require a balloon angioplasty.

The initial surgical procedure to establish flow into the true pulmonary arteries may be a shunt, a direct connection of the back of the aorta to the tiny central pulmonary arteries or an RV to PA conduit.

Intermediate stages – include catheter treatment of PA stenoses with angioplasty or stents, embolisation of aortopulmonary collaterals and surgical unifocalisation.

At the final operation, the VSD is closed - once the total cross-sectional area of the pulmonary vasculature is adequate to accept the full cardiac output i.e. enough segments of lung supplied from the RV via the true PA's without pulmonary hypertension, then the VSD can be closed with acceptable right ventricular pressures. The shunt is taken down, MPA is repaired and valved conduit between RV and the MPA is placed. However, if there are concerns with high RV pressure as a result of raised PVR or small pulmonary arteries, the VSD is fenestrated.

Specific postoperative problems
- LCOS or congestive cardiac failure
  Excess pulmonary blood flow as a result of dual supply from the newly created right ventricle to PA connection with persistent aortopulmonary connections can result in congestive cardiac failure. Excess L to R shunt can be treated by transcatheter embolisation/coil occlusion of aortopulmonary collaterals.

- Residual lesions –
  - Pulmonary outflow obstruction - may be amenable to dilation or stenting in the catheterisation laboratory. The risk of catastrophic vascular disruption can be significant in the immediate postoperative period.
  - Central pulmonary artery distortion will need surgical revision

- Left PA stenosis at the ductal insertion site in cases of TOF with PA where the duct is the source of the pulmonary blood flow. If a shunt is performed, close monitoring of the left lung perfusion by echo and or lung perfusion scan is important in the 1-4 months after surgery.

- Postoperative bleeding after final repair may be significant as a result of long operation; long suture lines and long period of pump suction.

- Right ventricular hypertension after VSD closure – may need revision with a fenestrated VSD patch to preserve adequate systemic blood flow.

- Cyanosis – infarction of segments of lung with subsequent infection and air leaks.
TETRALOGY OF FALLOT WITH ABSENT PULMONARY VALVE

TOF with absent pulmonary valve syndrome is clinically distinct from regular TOF. It is characterized by a large VSD, small pulmonary annulus, and nubbins of valve tissue instead of leaflets causing severe PS and regurgitation in utero. This results in massive dilatation of pulmonary arteries and compression of the airways. The cyanosis is from airway compression rather than right to left intracardiac shunting. In addition to the mediastinal abnormalities of the pulmonary arteries with bizarre branching patterns at the hilum; the intraparenchymal pulmonary vessels are abnormal as well with abnormal segmental arteries and elastic laminae. Finally, the airway itself is typically abnormal, with areas of tracheobronchomalacia and occasionally reduced numbers of bronchial generations.

The neonatal group presents typically within hours of birth with marked respiratory distress, cyanosis and air trapping due to tracheobronchial compression. The hypoxaemia is usually caused by a combination of right to left shunting at the VSD as well as pulmonary venous desaturation from ventilation perfusion mismatch. High positive end expiratory pressure PEEP may help to stent open floppy airways. Prone positioning is occasionally helpful in relieving some of the tracheal compression.

Early repair of this subgroup is necessary with anterior and posterior plication of the pulmonary arteries, closure of the VSD and transannular patching of the right ventricular outflow tract.

A new approach for correction of TOF with absent pulmonary valve has recently been described. This includes, in addition to the standard TOF repair, translocation of the pulmonary artery anterior to the aorta and away from the airways (Hraska et al, 2002). This technique has the potential to reduce or eliminate bronchial compression by pulmonary artery.

In general, despite a technically adequate repair, many of these symptomatic neonates have continued pulmonary difficulties. Nearly all have some degree of bronchomalacia in infancy and childhood and some require tracheostomy, long-term ventilation and PEEP.
TETRALOGY OF FALLOT WITH COMPLETE A-V CANAL

TOF, in combination with complete AVSD, poses significant surgical and management difficulties, which result in higher mortality rates and frequent postoperative residua. Surgical management of the RVOTO remains the same as for TOF.

**Important considerations** are to maintain competency of the pulmonary valve. Because the tricuspid component of the common A-V valve is frequently abnormal and regurgitant after repair, a transannular patch results in both free PR and TR and the combination typically results in severe right sided heart failure in the immediate postoperative period. In patients with severely deformed pulmonary valve or those with annular hypoplasia, valved conduits should be considered.

**Postoperative residual defects** should be sought including residual VSD, RV outflow tract obstruction, A-V valve regurgitation and conduction disturbances. Right-sided heart dysfunction is extremely common given the abnormalities of the right-sided A-V valve, infundibular incision and pulmonary regurgitation. Leaving a patent foramen ovale is particularly helpful in these children with borderline RV function.
TRICUSPID STENOSIS, HYPOPLASTIC RIGHT HEART SYNDROME AND PULMONARY ATRESIA WITH INTACT VENTRICULAR SEPTUM

The major pathophysiological abnormalities stem from right heart obstruction. The presence of pulmonary atresia means that these neonates have ductal dependant pulmonary outflow. Tricuspid stenosis requires the presence of an adequate atrial septal defect or venous hypertension (right atrial) will develop. The combination of pulmonary atresia with a patent but stenotic tricuspid valve forces the right ventricle to generate very high often suprasystemic pressures and dictates the presence of tricuspid regurgitation. In many of these patients, large connections develop between the hypertensive right ventricle and the coronary arteries (up to 29% in one large series). In children with pulmonary atresia, the presence or absence of VSD radically changes the anatomy, pathophysiology treatment and prognosis (Table 1).

Figure 7: PA + IVS

<table>
<thead>
<tr>
<th>Tricuspid valve</th>
<th>PA with VSD</th>
<th>PA without VSD</th>
</tr>
</thead>
<tbody>
<tr>
<td>ASD</td>
<td>Irrelevant</td>
<td>Necessary for survival</td>
</tr>
<tr>
<td>RV pressure</td>
<td>Equal to LV pressure</td>
<td>More than LV pressure</td>
</tr>
<tr>
<td>Large RV-coronary connections</td>
<td>Not found</td>
<td>Present, up to 29%</td>
</tr>
<tr>
<td>PA size</td>
<td>Often small</td>
<td>Normal</td>
</tr>
<tr>
<td>Pulmonary blood flow</td>
<td>Ductal dependent</td>
<td>Ductal dependent</td>
</tr>
<tr>
<td>Ultimate repair</td>
<td>Two ventricles</td>
<td>Fontan approach</td>
</tr>
</tbody>
</table>

Figures from http://images.md
Website resources provided by NHS England
In contrast to children with TOF who often have small pulmonary arteries, those with pulmonary atresia and intact ventricular septum (PA + IVS) have normal to generous sized pulmonary arteries. The key controllable determinant to right ventricular growth is pulsatile forward flow. This requires both a widely open outflow tract and an inflow i.e. the tricuspid valve then allows significant volume to enter the right ventricle. Without both of these factors, the right ventricle does not grow. The RVOT can be opened surgically but nothing can be done at present to enlarge the tricuspid valve. Data from the Congenital Heart Surgeons Study showed that the diameter of the tricuspid valve normalized to body surface area (tricuspid valve z-value) and was highly correlated with size of the right ventricular cavity. Therefore, tricuspid valve annulus size is used as a key predictive element to assess the likelihood of using the right ventricle as a pump in the repair operation or heading towards a single ventricle type approach.

Treatment of all neonates with pulmonary atresia with intact ventricular septum starts with PGE1 infusion. With stable, non-acidotic infants, one assesses all the factors to formulate a plan. **One must answer the following questions.**

1) Is the tricuspid valve too small to allow RV growth?
2) Does the ASD need to be enlarged?
3) What is the source of pulmonary blood flow, and how adequate is it?
4) Is obstruction to the systemic circulation present?
5) Are signs of RV/coronary artery connections present and are the native coronary arteries normal?

The final query can only be answered by selective RV angiography and aortography. Thus, all neonates with PA with IVS should have diagnostic cardiac catheterisation.

**Cardiac catheterisation**
- Allows right ventricular pressure measurement, confirms anatomic pulmonary atresia, and evaluates right and left ventricular function.
• Delineates ventriculocoronary connections and the morphology and size of the tricuspid valve and right ventricle.
• In the rare instance of a restrictive atrial communication, a transcatheter balloon or blade atrial septostomy may help maintain adequate cardiac output. Recently, transcatheter wire puncture, laser, and radiofrequency-assisted balloon pulmonary valvotomy have been utilized as alternatives to surgical valvotomy in patients with PA with IVS.

Ultimate repair options are two-ventricle repair, 1½ ventricle repair or 1 ventricle (Fontan approach).

The basic principles in surgical repair
• Avoid distortion of pulmonary arteries
• Minimise any tendency to increase medial muscle within pulmonary arterioles or to compromise pulmonary subsegments
• Ensure easy egress out of the lungs
• Maintain left ventricular compliance
Anything that increases the resistance from systemic veins to left ventricular ejection is harmful to patients designed for a Fontan type connection.

Blalock-Taussig Shunt
The first palliative step in treating infants with PA + IVS involves ensuring an adequate source of pulmonary blood flow. In neonates, this means a systemic to pulmonary artery shunt. When large right ventricle to coronary connections are present, surgical opening of the RV outflow tract may create a steal phenomenon from the coronaries into the RV thereby not perfusing the myocardium. These children require tricuspid valve closure.

Superior Cavopulmonary anastomosis
This involves surgical anastomosis of the transected end of SVC to the top of the right pulmonary artery leaving the two pulmonary arteries in free communication. Advantages are that it provides effective pulmonary blood flow provided the pulmonary arteries are of good size and unobstructed and the pulmonary resistance is low. Deoxygenated blood from the head, neck and upper body is delivered directly without admixture to the pulmonary circulation. It also unloads the left ventricle, reducing its workload and thereby its end diastolic pressure. It is an excellent second procedure for children with PA with IVS but not useful in neonates as a first procedure because of the unresolved pulmonary artery hypertension.

For complete separation of the two circulations, three options exist:-
• The two ventricle repair – available only to a few of these children because the tricuspid valve and the right ventricle are usually too small even after decompression of RV outflow tract obstruction and the growth is usually inadequate to achieve normal sized right heart structures.
  Right ventricular overhaul procedure could be performed in some substrates by recruiting more functional right ventricle by division of apical and outflow trabeculations. Right ventricular diastolic dysfunction, tricuspid and pulmonary regurgitation and atrial arrhythmias tend to remain as long term problems.
• Fontan variant – it combines direct cavopulmonary anastomosis with intra-atrial tunnel through the right atrium to connect IVC to pulmonary artery. As the tricuspid valve is closed, excluding the RV from the circulation, ASD closure is not necessary. Functionally, these children are acyanotic (in absence of fenestration or collaterals) and have reasonable exercise tolerance but can only increase cardiac output to a maximum of 2-3 times basal whereas normal children might increase cardiac output as much as 7 fold during strenuous exertion.
• 1½ ventricle approach – this is applicable to those whose tricuspid valve and right ventricle can handle approximately 60% of the venous return. The SVC generally transmits 40%. This 1½ ventricle repair involves a superior vena cavopulmonary anastomosis combined with ASD closure, allowing IVC blood to flow to the RA and through the tricuspid valve or the right ventricle and into the lungs to the surgically opened RV outflow tract. Some implant a pulmonary valve but most do not because this places a fixed element in the growing heart.
**Flowchart for management options in PA+IVS**

**Predictors of successful biventricular repair** are greater preoperative weight, higher tricuspid valve z-scores, and tricuspid/mitral valve ratios. A tricuspid/mitral ratio >0.5 has been reported to be the best predictor of a biventricular repair.

The moderate term outlook 10-15 years for children with 1 ventricle repair is good but long-term problems with progressive venous hypertension, protein losing enteropathy, arrhythmia and pulmonary thrombosis are reported.

**POSTOPERATIVE ISSUES IN PULMONARY ATRESIA WITH IVS**
- Low cardiac output state in the immediate postoperative period
- One of the possibilities for LCOS includes unrecognised RV dependant coronary circulation with myocardial ischaemia. ECG changes, ventricular arrhythmias and segmental wall akinesis or dyskinesis can be seen on echocardiography. Large areas of myocardial ischaemia following this procedure carry a grave prognosis.

- A second form of LCOS can develop 1-3 days following surgery resulting from a circular shunt. When the repair involves placement of systemic to pulmonary artery shunt and a transannular right ventricular outflow tract patch, an ineffective circular shunt becomes possible. The systemic to pulmonary artery connection in combination with the obligatory pulmonary insufficiency, associated tricuspid valve insufficiency and atrial septal defect can result in a circular flow pattern as described below. Blood from the left atrium into the left ventricle and the aorta, then passes through the systemic to pulmonary artery shunt into the pulmonary arteries, then retrograde across the RV outflow tract (by design, the transannular patch results in pulmonary regurgitation) in diastole and retrograde across the tricuspid valve into the right atrium in systole and then across the right atrium back into the left atrium. This results in an inadequate delivery of effective systemic blood flow. Patients with a large circular shunt can develop oliguria, metabolic acidosis and systemic hypotension. Manoeuvres to increase pulmonary vascular resistance and lower systemic vascular resistance may be helpful. Surgical manoeuvres to narrow the shunt and or minimise the TR may be necessary.

- LCOS due to small RV, RV ventriculotomy, severe pulmonary regurgitation.

- A large PDA not clipped at surgery, or which remains open after stopping PGE can contribute to a systemic steal syndrome and cause a LCOS.

- Significant persistent hypoxaemia – seen in patients with residual RV outflow tract obstruction or severe tricuspid hypoplasia. Exclude a pulmonary cause.

- High RA pressures due to small, non-compliant RV may cause ascites, pleural and pericardial effusions.

**Cardiac echo** to look at RV function – systolic and diastolic, pulmonary regurgitation, intracardiac shunt and pressure gradient across pulmonary valve and infundibulum should be done. Higher systolic blood pressure may be necessary and noradrenaline may be preferable to beta agonists, which have vasodilator properties. Avoid vasodilators.
TRICUSPID ATRESIA

Tricuspid atresia is a condition in which there is no direct patent communication between right atrium and right ventricle. The uniform pathophysiologic abnormality is an obligatory total right to left atrial shunt. Additional physiological consequences are dependent on the presence, severity and/or size of 3 factors: 1) a VSD 2) pulmonary stenosis 3) great artery relations.

When tricuspid atresia (TA) and no VSD are present, no flow occurs in utero to induce the development of a right ventricle. Thus, TA without a VSD means an absent RV. If a VSD is present, its size determines the left to right ventricular flow and thus there is a growth stimulus to the right ventricular cavity and with a large VSD, there can be a nearly normal sized right ventricle.

These children can have mild, moderate or severe RV outflow tract obstruction at the level of the valve. When the right ventricle is small, the VSD and the RV cavity together obstruct blood flow into the outflow area. When the VSD and right ventricle are large, the degree of RV outflow tract obstruction is variable.

Finally, the great arteries may be normally related great arteries or be transposed. With transposition and RV outflow tract obstruction with or without a small RV or a small VSD, these children functionally have hypoplastic left heart syndrome.

Types of Tricuspid Atresia – Classification

This unified classification includes all the previously described abnormalities in the positions of the great arteries.

Table 2. Classification of Tricuspid Atresia

<table>
<thead>
<tr>
<th>Type I</th>
<th>Type II</th>
<th>Type III - Great artery positional abnormalities other than D-transposition of the great arteries</th>
<th>Type IV</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>Subtype 1</td>
<td>Subtype 2</td>
</tr>
<tr>
<td>Normally related great arteries</td>
<td>D-Transposition of the great arteries</td>
<td>L-Transposition of the great arteries</td>
<td>Double outlet right ventricle</td>
</tr>
</tbody>
</table>

Subgroup a: Pulmonary atresia

Subgroup b: Pulmonary stenosis or hypoplasia

Subgroup c: No pulmonary stenosis

Management - the objective is not only to provide symptomatic relief but also to preserve, protect, and restore anatomy (good-sized and undistorted pulmonary arteries) and physiology (normal pulmonary artery pressure and preserved left ventricular function) to normal so that a final procedure can be safely performed when the patient reaches an optimal age and weight.

- In the neonate, obstruction at the level of the atrial septum may be treated with conventional Rashkind balloon atrial septostomy
- In most patients, obstruction to pulmonary blood flow is at the VSD level or in the subpulmonary region. In some patients, the obstruction is at the pulmonary valve. In these patients, balloon pulmonary valvuloplasty may be useful in improving pulmonary blood flow and oxygen saturation.

Surgical Management
Great Ormond Street Hospital ITU modular training programme

- **Modified Blalock-Taussig shunt** if decreased pulmonary blood flow - Pulmonary blood flow may be increased by surgical creation of an aortopulmonary shunt.
- **PA banding** if increased pulmonary blood flow - Patients with increased pulmonary blood flow are likely to have type Ic or type IIC defects without associated pulmonary stenosis. Congestive heart failure is likely to occur in these patients.
- In patients with **tricuspid atresia type II (TGA)**, PA banding should be performed following stabilization with anti-heart failure measures. Banding not only improves congestive heart failure but also helps achieve normal pulmonary artery pressure so that bidirectional Glenn and Fontan procedures can be safely performed later. If associated aortic co-arctation is present, it must be relieved. Pulmonary artery banding stimulates more ventricular hypertrophy, which may further reduce the size of the VSD, thus increasing subaortic obstruction.
- In patients with **tricuspid atresia type I (normally related great arteries)**, aggressive anti-heart failure measures should be promptly undertaken. Pulmonary artery banding generally is **not recommended** in this group of patients as the VSD closes or becomes smaller with time and the patients with pulmonary plethora develop pulmonary oligemia.
- In patients with **intracardiac obstruction** (may occur at the level of PFO and VSD).
- **Balloon/Blade atrial septostomy** if interatrial obstruction - Because the entire systemic venous return must pass through the patent foramen ovale, it should be large enough to allow unimpeded egress of systemic venous blood. Blade atrial septostomy is occasionally necessary, especially in older infants and children. Surgical atrial septostomy is needed even less frequently. However, it may be performed concurrently with bidirectional Glenn procedure.
- **Interventricular obstruction**: Spontaneous closure of the VSD can occur, causing interventricular obstruction.
  - Functional VSD closure in **tricuspid atresia type I** results in cyanotic spells, similar to those in TOF. If no improvement occurs in response to conventional treatment, emergency surgical palliation with a Blalock-Taussig type of shunt may be necessary.
  - In patients with **tricuspid atresia type II**, spontaneous closure of the VSD produces subaortic obstruction. This obstruction should be relieved or bypassed as soon as it is detected because it produces left ventricular hypertrophy, which in turn increases the risk at the time of Fontan operation. The VSD, right ventricle, and aortic valve may be bypassed by anastomosis of the proximal stump of the divided pulmonary artery to the ascending aorta (**Damus-Kaye-Stansel procedure**) directly or via a prosthetic conduit at the time of bidirectional Glenn or Fontan conversion. Alternatively, the conal septal muscle may be resected, enlarging the VSD; this is a direct approach in relieving the subaortic obstruction. However, development of heart block, inadequate relief of obstruction, and spontaneous closure of the surgically enlarged/created VSD remain major concerns.

**Completion of single ventricle palliation with**

- Bidirectional cavopulmonary anastomosis
- TCPC/Fenestrated Fontan

**Current surgical approaches**

- The patient's age, weight, and anatomic and physiologic status determine the types of surgery recommended. The overall objective is to achieve a total cavopulmonary connection.
- In the **neonates and young infants with pulmonary oligemia**, classic or modified Blalock-Taussig shunting is undertaken to improve the pulmonary oligemia.
- In **patients aged 6 months to 1 year**, Blalock-Taussig shunting and bidirectional Glenn procedure are the choices.
- For **children aged 1-2 years**, the bidirectional Glenn procedure is preferable.
For patients older than 2 years, total cavopulmonary connection may be performed, but most authorities suggest staging by using an initial bidirectional Glenn operation followed by Fontan conversion in 6-12 months.

Important issues

- At the time of bidirectional Glenn surgery, any narrowing of the pulmonary artery should be repaired. Issues related to subaortic obstruction and mitral valve regurgitation should also be addressed.
- Before Fontan conversion, cardiac catheterization should be undertaken to ensure normal anatomy and pressure of the pulmonary artery as well as normal LVEDP. At the same time, aortopulmonary collaterals should be evaluated by means of selective subclavian artery and descending thoracic aortic angiography. If collateral vessels are present, they should be occluded with coils. Some authors question routine use of pre-Fontan catheterization and suggest prospective evaluation of this issue.
- At the time of Fontan conversion, most surgeons currently prefer extracardiac conduit diversion of inferior vena caval blood into the right pulmonary artery. To address the growth issue related to extracardiac Fontan surgery, some surgeons use autologous pericardial roll grafts.
- In patients with associated transposition of the great arteries, early banding of the pulmonary artery, relief of aortic coarctation (if present), and bypassing (by means of a Damus-Kaye-Stansel procedure) or resecting the subaortic obstruction should be incorporated into the management plan.

http://www.emedicine.com/ped/topic2550.htm
EBSTEIN'S ANOMALY

Ebstein's anomaly refers to a malformation of the tricuspid valve that can be so mild that it is found incidentally at autopsy or so severe that it causes fetal demise. There is a continuum of severity related to how displaced tricuspid valve orifice is into the right ventricular cavity.


| Top, Normal | Tricuspid valve with anterior, posterior, and septal leaflets in 1 plane. |
| Middle, Right-sided | Ebstein's anomaly showing displacement of posterior and septal leaflets; maximal displacement is at the crux of the posterior and septal leaflets. |
| Bottom, Left-sided | Tricuspid valve in left-sided Ebstein's anomaly; the displacement of leaflets is similar to that in the right-sided anomaly. |

Picture from Article by Jost Ebsteins anamoly Circulation 2007.

The components of an atrioventricular valve include the annulus, a fibrous ring on which the leaflets attached to provide the hinge point on which the leaflets move, the orifice where the leaflet edges co-apt and through which blood flow passes, leaflets which funnel the blood in ventricular diastole but prevent flow in ventricular systole, chordae which support the leaflets during systole and through which the blood actually enters the ventricle and papillary muscles which in concert with the adjacent myocardium contract during systole to draw the chordae to the proper length and tension for optimal valve function.

In patients with Ebstein's anomaly, the posterior and septal leaflets are adherent to the myocardium, thereby displacing the orifice of the valve downward into the right ventricle cavity away from the annulus. As the displacement is more pronounced, the physiologic consequences increase in magnitude. The tricuspid valve becomes increasingly regurgitant and this can be sufficiently severe to reduce cardiac output. Encroachment on RV cavity size reduces net cardiac output as well as forward flow to the RV outflow tract so as to cause pulmonary stenosis or even atresia. The atrialised right ventricle causes discoordination of flow and contraction that interrupts the normal directional blood flow pattern within the heart and results in functional stenosis.

Associated problems include 1) ASD 2) pulmonary stenosis or atresia 3) hypoplasia of pulmonary arteries 4) PDA. These infants often have arrhythmias from the extreme enlargement of the atrium and due to the frequent associated occurrence of the Wolff Parkinson White syndrome.

Treatment
Medical - includes prostaglandin infusion for maintenance of the duct, pharmacological therapy for any rhythm problems and afterload reduction to encourage forward flow.

Surgical
There are no good surgical options for neonates with Ebstein's anomaly. Those who require an operation as neonates generally do not survive; surgical closure of the tricuspid valve together with a small aortopulmonary shunt has been used with limited success.
Surgical ablation procedures for accessory pathway-mediated tachycardia and atrioventricular nodal re-entrant tachycardia have been reported to give excellent freedom from recurrence of arrhythmias.

Cardiac transplantation might be contemplated but pulmonary hypoplasia may preclude the success of any intervention short of heart/lung transplantation.

Patients who survive the infant period and require later surgery can be treated with reconstructive procedures that plicate the atrialised right ventricle in conjunction with the annuloplasty of the tricuspid valve. In older patients, tricuspid valve replacement is seldom necessary.


| Diagram of the tricuspid valve repair technique |
| A, Two papillary muscles arise from the free wall of the right ventricle, with short chordal attachments to the leading edge of the anterior leaflet. The septal leaflet is diminutive and only a ridge of tissue. The posterior leaflet is not well formed and is adherent to the underlying endocardium. A small patent foramen ovale is present. |
| B, C, The base of each papillary muscle is moved toward the ventricular septum at the appropriate level with horizontal mattress sutures backed with felt pledgets. The patent foramen ovale is closed by direct suture. |
| D, The posterior angle of the tricuspid orifice is closed by bringing the right side of the anterior leaflet down to the septum and plicating the nonfunctional posterior leaflet in the process. |
| E, A posterior annuloplasty is performed to narrow the diameter of the tricuspid annulus. The coronary sinus marks the posterior and leftward extent of the annuloplasty. |
| F, An anterior purse-string annuloplasty is performed to further narrow the tricuspid annulus. This annuloplasty stitch is tied down over a 25-mm valve sizer in an adult to prevent tricuspid stenosis. |
| G, Completed repair that allows the anterior leaflet to function as a monocuspid valve. |
Other sources of information:
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