Pre- and Post-operative Management of Cardiac Shunt Lesions

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Information for Year 1 ITU Training (basic):

Year 1 ITU curriculum
- General anatomical concepts: PDA, ASD, VSD, AVSD.
- General surgical repair techniques
- Post operative ICU considerations

Curriculum Notes for Year 1:

ATRIAL SEPTAL DEFECTS –
Figure 1: Types of ASD

[Diagram with labels]

CS – coronary sinus
IVC – inferior vena cava
OP – ostium primum
OS – ostium secundum
RA – right atrium
RAA – right atrial appendage
RV – right ventricle
SV – sinus venosus
SVC – superior vena cava

Figures from http://images.md
Website resources provided by NHS England
PRE-OPERATIVE ANATOMICAL AND CLINICAL CONSIDERATIONS

- All ASDs result in a high pulmonary blood flow situation with a L to R shunt leading to a volume overload of RA, RV, PA and LA. The degree of L to R shunt depends on the size of the ASD as well as the relative compliance of the ventricles during diastole. Blood flow across predominantly during diastole when the mitral and tricuspid valves are open and filling occurs in the ventricles.
- Ostium primum ASDs have associated abnormalities of AV valves – most commonly a cleft in the septal leaflet of the mitral valve.
- Sinus Venosus ASDs – there are 2 subtypes – the high SVC type occurs in the posterosuperior aspect of the atrial septum and is associated with partial anomalous drainage of the right upper and middle pulmonary vein. This vein can drain into the SVC or the RA. The second type is the IVC type, which is situated low and can also be associated with PAPVD.
- Coronary Sinus ASD is uncommon where the hole is present in the roof of the coronary sinus and hence the coronary sinus and left atrium are in continuity and in communication with the RA. This can also be associated with a persistent left SVC.

SPECIFIC POSTOPERATIVE PROBLEMS
Repair of ASD is usually uneventful. Aim for early extubation.

Sino-atrial dysfunction – can occur as a result of either direct trauma or as a consequence of interruption of blood supply. The incidence is higher in the sinus venosus type ASD repair. Close monitoring of the heart rhythm is important, as the rhythm disturbance may be subtle with inappropriate chronotropic response to anaemia, stress and inotropic agents.

Postpericardiotomy syndrome – any fever and systemic signs should be investigated with CXR and Echo. Pericardial effusion if present with associated haemodynamic compromise needs to be urgently drained.

AV block – Seen after repair of the coronary sinus ASD as a result of close proximity of the defect to the AV node.

LV dysfunction and pulmonary hypertension - seen in older patients with chronic right ventricular overload and preoperative pulmonary vascular hypertension respectively are uncommon in the paediatric population.

The risk of death for patients undergoing surgical closure of uncomplicated ASD is less than 1%.

VENTRICULAR SEPTAL DEFECTS

PRE-OPERATIVE ANATOMICAL AND CLINICAL CONSIDERATIONS

Classification of Ventricular Septal Defects (VSD)
Classification of ventricular septal defects (VSDs) is based on the relation of the VSD to the membranous septum (MS) and the tricuspid and arterial valves.
The interventricular septum as viewed from the RV to indicate the location and classification of VSD. 1: Inlet defect; 2: perimembranous defect; 3-5: muscular/trabecular defect; 6-7: subarterial defect.

**Perimembranous (PM) defects** are those that involve the interventricular membranous septum and the adjacent muscular septum. This places the VSD in the outflow tract of the left ventricle and immediately under the aortic valve. The aortic and tricuspid valves are in direct contact through the defect. The mitral valve is separated from the defect by the intervening atrioventricular membranous septum. The atrioventricular conduction axis is intimately related to the posteroinferior margin of the perimembranous defect. A PM VSD can close spontaneously by apposition of the septal leaflet of the tricuspid valve to the defect. These defects can extend toward the inlet, the trabecular part, or the outlet of the right ventricle.

**Subarterial (subpulmonary or supracristal) VSD** is located above the crista supraventricularis, which places it in the RV outflow tract and immediately below the right cusp of the aortic valve. The aortic and pulmonary valves are in direct contact at the roof of the defect. These defects are frequently associated with the prolapse of the right aortic cusp and resultant aortic insufficiency. Once this occurs the VSD needs to be repaired as soon as possible to avoid permanent damage to the aortic valve cusp.

**Inlet VSD** is located posteriorly just inferior to the tricuspid and mitral valves. This is a type of endocardial cushion defect and does not close spontaneously.

**A muscular VSD** is located in the muscular interventricular septum and can be multiple and have the highest rate of spontaneous closure.

**Defects can involve more than one of the septal components and are called perimembranous confluent defects.**

The amount of pulmonary blood flow to as compared to systemic blood flow is called the Qp/Qs ratio.

**Size classification of VSD’s**
- Based on the maximum measured diameter of the defect as compared to a normal aortic valve annulus
Small VSD: ≤ 1/3 of the diameter of the aortic valve annulus
Moderate sized VSD: about ½ of the diameter of the aortic valve
Large VSD: near the same diameter of the aortic valve annulus

- Clinically based on Qp/Qs ratio
  - Small VSD - Qp-Qs ratio < 1.5
  - Moderate sized VSD - Qp-Qs ratio 1.5 – 2.0
  - Large VSD - Qp-Qs ratio > 2.0

The degree of shunt is determined by the size of the defect and respective vascular resistances (PVR and SVR). Ventricular level shunting takes place predominantly during systole when the AV valves are closed and the semilunar valves are open.

Associated Lesions
- PDA, ASD, pulmonary stenosis (DORV), subaortic stenosis, and coarctation of aorta

Malaligned defects
Malaligned defects are characterised by malalignment of the conal septum and trabecular muscular septum and occur in a similar position to perimembranous defects and typically occur as a component a more complex cardiac defect.

Congestive cardiac failure
Infants with large VSDs may have significant congestive heart failure and failure to thrive and be on large doses of diuretics, digoxin, captopril and high caloric formula.

Surgical technique
The approach is through a right atriotomy for PM VSD, Inlet VSD and muscular VSDs and Pulmonary arteriotomy or small horizontal infundibulotomy for outlet VSD. In the repair of PM VSD, care must be taken to avoid the aortic valve annulus and aortic valve (which is close to the superior rim of the defect) and the conduction system (which is close to the posterior and inferior rim of the defect).

SPECIFIC POSTOPERATIVE PROBLEMS

Residual VSD
A residual VSD has to be investigated for if there is low cardiac output state, persistent metabolic acidosis, pulmonary oedema, high PA and LA pressures. A significant residual VSD will need re-operation.

Pulmonary hypertension
Postoperative pulmonary hypertension may be caused by residual VSD, postoperative changes in pulmonary vascular resistance or undiagnosed branch pulmonary artery stenosis. Episodic elevation of PA pressures suggests postoperative pulmonary hypertension.

Heart Block
Postoperative heart block can occur in up to 10% of patients after VSD closure and is more common after perimembranous type VSD, outlet and muscular VSD repair.

Junctional Ectopic Tachycardia: Post operative

Etiology
- JET is thought to arise from abnormal automaticity of the AV node or proximal His bundle due to direct injury/ stretch, infiltrative haemorrhage, ischemia or oedema.
- Histamine, eosinophil cation protein, or other products of mast cell, eosinophil, or basophil degranulation that are liberated in response to cardiopulmonary bypass have been implicated in the genesis of transient postoperative JET.
- JET is driven by a focus within or immediately adjacent to the atrioventricular (AV) junction of the cardiac conduction system (i.e., AV node–His bundle complex), but does not have the features associated with re-entrant tachycardia (e.g., AV node re-entry).
Incidence/ Timing
- More commonly seen in patients less than 1 year of age
- Incidence anywhere from 5% to 20%; higher incidence after certain types of CHD repairs. See Risk Factors.
- JET usually begins 6-72 hours following cardiopulmonary bypass.

Defining Characteristics/ Diagnosis
- JET is classically recognized as a narrow QRS tachycardia (160-260/min) with AV dissociation, and an atrial rate, which is slower than the ventricular rate.
- The ectopic focus emits impulses at a rapid rate, which are conducted 1:1 down the His-Purkinje system. At the same time the atrium is activated by the normal sinus impulses, at a much slower rate. Thus, there is AV dissociation, except for sinus capture beats resulting from occasional antegrade conduction of a normal sinus impulse, causing the next QRS complex to occur slightly earlier than expected. Such capture beats, since they utilize the same His-Purkinje system as the junctional focus; will occur with an identical QRS morphology to that seen with the underlying JET.
- In young patients, the AV node has the capacity to conduct rapid junctional rates in a retrograde fashion (particularly in the presence of inotropic agents) and AV dissociation therefore may not be present.
- The tachycardia does not respond to a single extrastimulus and does not convert with programmed stimulation or cardioversion.
- The use of atrial wire recordings to assess P wave timing can facilitate determination of the diagnosis. In some patients with 1:1 AV or VA association, whether the rhythm is being driven by the atrium or junction may be unclear. If this occurs, pacing the atrium faster than the intrinsic rhythm and then identifying the origin of the first escape beats following termination of pacing may be helpful.
- Administration of adenosine can result in loss of VA conduction with perpetuation of JET.
- An atrial premature beat introduced immediately before the expected atrial timing would not conduct retrogradely if the origin of the intrinsic atrial depolarization were from retrograde conduction of JET, whereas sinus rhythm with first-degree AV block would respond by advancing the atrium and the ventricle.
- The 2 ECG patterns typically observed are
  - Junctional rhythm with 1:1 retrograde VA conduction
  - Junctional rhythm with retrograde VA dissociation
- In the latter case, the ventricular rhythm is irregular, with an early ventricular depolarization occurring whenever the dissociated P wave occurs with a timing that can conduct in the antegrade direction. An exception to these patterns occurs rarely, when both JET and complete heart block are present.
- Another exception to the classic ECG appearance is the occurrence of bundle branch block and the QRS may appear wide. This may be rate-dependent aberration, due simply to the rapid rates, or may be fixed and due to surgical damage to the bundle branch. An example of the latter is the child who develops post-operative JET after repair of Tetralogy of Fallot, which is often associated with a right bundle branch block. Differentiating JET from ventricular tachycardia is difficult in the face of bundle branch block.

Risk Factors
- JET occurs more frequently in certain types of CHD repairs - TOF, VSD, AVSD
- Other operative risk factors for developing JET
  - Resection of muscle bundles, higher bypass temperatures and relief of right ventricular outflow tract obstruction through the right atrium
  - Relief of RVOT obstruction appeared to be more important in the causation of JET than VSD closure.
  - Muscular resection seems to be more arrhythmogenic than is simple division.
In another study of 33 patients with JET, univariate analysis revealed that dopamine or milrinone use postoperatively, longer cardiopulmonary bypass times, and younger age were associated with JET. Multivariate modeling elicited that dopamine use postoperatively (odds ratio, 6.2; \( p = 0.01 \)) and age less than 6 months (odds ratio, 4.0; \( p = 0.02 \)) were associated with JET.


**Treatment**

- As JET is transient in nature, temporizing measures are key.
- If the patient can tolerate the JET and/or one controls it with medication if hemodynamic compromise coexists, it seems to disappear in a few days of 'healing.'
- JET usually occurs at an extremely vulnerable period following cardiac surgery, when ventricular function is often diminished. The additional insults of poor ventricular filling because of tachycardia and the loss of AV sequential contraction are considered to contribute significantly to morbidity and mortality.
- Despite the above statement, not all patients "suffer" from their JET, in that good hemodynamics are maintained. If the patient is compromised, it is not subtle (sudden severe decreased cardiac output resulting in hypotension). However, if AV synchrony is restored, the cardiac output and blood pressure will increase just as immediately and dramatically.
- Treatment is aimed at slowing junctional rate to physiologic range and restore AV synchrony.
- Other general measures should be undertaken such as increasing sedation, decreasing catecholamines (they can be arrhythmogenic), optimizing preload, correcting electrolytes (particularly magnesium), and inducing controlled hypothermia (34- 35°C).
- Magnesium has been evaluated in a prospective double-blind placebo controlled study, in which either magnesium or normal saline was given immediately post cardiopulmonary bypass. After 28 children had been randomized, the study was terminated due to 27% of the placebo patients developing JET and none of the patients who received magnesium patients.

**Dorman, B. Hugh PhD .** American Heart Journal 2000:139(3):522-528

- Postoperative JET has been successfully controlled with amiodarone, propafenone, procainamide, or flecainide. Propranolol or sotalol have also been used in the therapy of these rhythm disorders.
- Amiodarone inhibits AV conduction and sinus node function. Prolongs action potential and refractory period in myocardium and inhibits adrenergic stimulation.
- Once the JET rate is reduced, the use of atrial or AV sequential pacing can help to restore AV sequence and cardiac output.
- Occasionally, atrial high-rate pacing to the point of 2:1 AV block can provide a controlled ventricular response while continuing to suppress the JET focus. This finding suggests a relatively high insertion site of the JET focus into the AV conduction system.
- The primary functions of surgical care in postoperative JET are to correct major residual defects that may be contributing to morbidity, to ensure that atrial-based pacing can be achieved, and to provide extracorporeal life support (i.e., extracorporeal membrane oxygenation [ECMO]) if required.
- In patients with postoperative JET, the presence of significant postoperative residual hemodynamic abnormalities should be excluded by transthoracic echocardiography, transesophageal echocardiography, or cardiac catheterization, if necessary.
Post operative JET ECG
6 month old child with JET post AVSD repair
ATRIOVENTRICULAR CANAL DEFECT OR ENDOCARDIAL CUSHION DEFECTS

Incomplete (Partial) AVSD includes ostium primum ASD, cleft mitral valve and defects of the atrioventricular septum (Gerbode defect). There is no interventricular communication and two separate AV valves are present.

Complete AVSD – the inferior portion of the atrial septum and the posterior portion of the ventricular septum are absent associated with one large common atrioventricular valve.

Transitional AV canal defect – An ostium primum defect is present and the atrioventricular valves may only be partially separated into two valves. Interventricular communication may small or moderate.

Figure 3. The level of communication possible in AVSDs
A, A purely atrial level of communication - the AV valves are adherent to the ventricular septum, with no communication at the ventricular level.
B, Only ventricular communication. The AV valves have fused with the lower edge of the atrial septum, obliterating the ostium primum defect, and only the interventricular level shunt is present here. This defect is often called a ventricular septal defect of the AV canal type.
C, Combined levels of communication above and below the AV valves.

From [http://images.md](http://images.md)
Website resources provided by NHS England

Ventricular hypoplasia of either the right or left ventricle may exist and can preclude a biventricular repair.

Figure 4: Diagram demonstrating the balance of the AV orifice to the underlying ventricular mass
Diagram demonstrating the "balance" of the AV orifice to the underlying ventricular mass. A, The usual condition with a common AV valve orifice overlying both ventricles. B, Right-dominant AVSD. C, Left-dominant AVSD with the orifice situated predominantly over the right ventricle (RV). The right and left parts of the AV junction are usually committed evenly to the respective ventricles, but unbalanced commitment to either ventricle may occur, resulting in RV or left ventricular (LV) dominance. In most situations the AV junction is connected to the ventricular mass so that its left and right parts drain more or less evenly into both ventricles. If the AV junction is connected predominantly to either one or the other ventricle, then LV or RV dominance is said to exist seen in approximately 5% of the cases. More are of the right-dominant variety than of the left-dominant variety.
Figure 5: The classification of complete AVSDs

The classification of complete AVSDs, popularized by Rastelli et al., is based on the morphology of the anterosuperior bridging leaflet (ASBL), which can be displayed exquisitely by echocardiography. This diagram of a complete AVSD depicts the types of AV valve attachments of the ASBL that are identifiable echocardiographically and fit the surgical description of Rastelli. The left-hand panels are four-chamber views, while the right-hand panels are equivalent to subcostal short-axis or parasternal short-axis views.

A, Rastelli type A defect. Here the ASBL can be seen to be attached to the papillary muscle (PM) lying on the crest of the septum between the left (LV) and right (RV) ventricles. In the subcostal view, the posteroinferior bridging leaflet (PIBL) is depicted to be attached to the crest of the septum.

B, Rastelli type B defect. The ASBL is not attached to the septum, but to PM arising from the RV. C, Rastelli type C defect. The ASBL is attached to PM, which also supports the other leaflet of the tricuspid valve yielding a free-floating nonattached leaflet. The arrows in the right-hand panels indicate how the PM attachment is from a septally attached (type A) to RV-originating PM (type B), and PM fused with the anterior PM within the RV (type C).
Surgical Principles

- Examination of AV valves – papillary muscle, chordal attachments and ventricular size
- Zone of apposition of LAVV – to close or not; contra-indications to closure are the presence of small mural leaflet, parachute or single papillary muscle, and double orifice LAVV
- Creation of two non stenotic valves
- Closure of VSD and ASD with patch – Different techniques available – single patch, two patch and modified single patch
- Avoidance of the conduction system

AVSD – SPECIFIC POSTOPERATIVE PROBLEMS IN ICU

Low Cardiac Output State
Residual AV Valve disease
Pulmonary Hypertension
Arrhythmias
Downs Syndrome
Associated Cardiac Lesions

Low Cardiac Output State

Fall in cardiac output after congenital open-heart surgery is seen in the immediate postoperative period. The causes are multifactorial and contributes to postoperative mortality and morbidity. Myocardial Dysfunction in the postoperative period may be caused by the
inflammatory response after CPB, myocardial ischaemia after aortic crossclamp, hypothermia, re-perfusion injury and inadequate myocardial protection.

**Causes of LCOS in the postoperative AVSD**
- Dehiscence of the repair of the left AV valve
- Residual VSD
- Residual LAVVR
- Poor LV function
- LVOTO

Assess LCOS by clinical parameters - HR/rhythm, blood pressure, filling pressures – CVP/LA, peripheral perfusion and temperature, urine output and any alteration in CNS state as well as by laboratory parameters such as acid base balance, mixed venous oxygen saturation (SvO₂) and lactate.

**Causes of elevated left atrial pressure after AVSD repair**
- Left AV valve regurgitation
- Left AV valve stenosis
- Ventricular outflow tract obstruction
- Residual VSD
- Left ventricular dysfunction
- Pericardial effusion/tamponade
- AV dyssynchrony

**Postoperative LAVVR**
The incidence of re-operation for LAVVR is high – 5-10%. It is related to the anatomy of the AV valve, age at repair, surgical techniques employed and is an associated risk factor for postoperative death (Michielon et al. Ann Thorac Surg 1995;60:604-609). Pre-operative LAVVR and double orifice ‘LAVV’ were associated with increased risk of postoperative LAVVR. Severe left AV valve regurgitation causes further annular dilatation, which in turn further aggravates LAVVR This vicious cycle of annular dilatation results in further low cardiac output and hypotension. Aggressive afterload reduction is mandatory and in a haemodynamically unstable patent, may necessitate a re-operation without delay.

**Pulmonary Hypertension** – The incidence is high in the immediate postoperative period and can contribute to postoperative mortality. It is seen more commonly in older patients at operation and in patients with Down’s syndrome. Microemboli, atelectasis, hypoxic pulmonary vasoconstriction, inflammatory mediators, endothelial injury have been implicated in the cause of post CPB pulmonary hypertension. Down’s Syndrome is an independent variable in the development of pulmonary vascular disease. Morphometric studies of lungs in patients with Trisomy 21 have shown inadequate alveolarisation in terminal lung units and comparable reduction in total cross sectional area of the vascular bed. Also, presence of upper airway obstruction, chronic hypoventilation and hypoxemia has been implicated in the increased pulmonary vascular resistance in this group of patients.

**Pulmonary hypertensive crisis** are identified by haemodynamic changes with an increase in CVP, fall in LA pressure, drop in oxygen saturations and fall in systemic blood pressure and a clinical impression of stiff lung. Changes in pulmonary vasculature may affect lung mechanics with a fall in lung compliance as the PA pressure increases. It is important to rule out anatomic causes of elevated PA pressure such as severe left AV valve regurgitation, left AV valve stenosis and residual VSD.

**Management principles** include moderate hyperventilation, moderate alkalosis, adequate inspired oxygen, normal lung volumes, optimal haematocrit, inotropic support, pulmonary vasodilators – iNO, avoidance of pain and ensuring adequate sedation

**Inhaled Nitric Oxide** is a selective potent pulmonary vasodilator with no systemic vasodilatation. It has a rapid onset and improves intrapulmonary shunt and reduces RV afterload. It has to be used with caution with patients with severe left ventricular dysfunction. On abrupt withdrawal, rebound pulmonary hypertension has been reported.
Arrhythmias after AVSD repair

- **Complete Heart Block**
  The postoperative incidence of complete heart block is around 4 to 5%. AV sequential pacing may be needed and consider permanent pacemaker if CHB persists past post operative day 10.
- **Sino-atrial node dysfunction**
  This is seen due to the abnormal position of the AV node as a result of lack of the AV septal structures
- **RBBB**
- **JET** – diagnosis and treatment as described above

Residual VSD

**The clinical signs**

- Sinus tachycardia
- Elevated PA and LA pressures
- Persistent metabolic acidosis
- Big heart and wet lungs - pulmonary oedema and cardiomegaly
- High ventilatory requirements
- Failure to wean from ventilator

**Management** includes inotropic support, vasodilators, diuretics and mechanical ventilation, which reduces LV afterload and decreases preload. If conventional management fails, re-operation may be needed.

Associated lesions

The presence of associated lesions such as Tetralogy of Fallot, Pulmonary stenosis, small restrictive ventricle can increase complexity of the postoperative course.

**Principles of ICU management after AVSD repair**

- Maintain cardiac output with vasodilators and inotropes
- Avoid rapid volume loading to prevent annular dilatation and increased LAVVR
- Anticipate pulmonary hypertension
- Watch for arrhythmias
- If deteriorating CO, suspect LAVVR, residual VSD

References.

- Aspects of paediatric cardiopulmonary bypass – Alex Robertson Department of Perfusion Great Ormond Street Hospital NHS Trust
- Pediatric Cardiac Intensive Care – Eds A Chang, F Hanley, G Wernovsky, D Wessel - Pre-operative Care of CHD

Websites

- [http://pedsccm.wustl.edu/All-Net/english/cardpage/operate/bypass/intro.htm](http://pedsccm.wustl.edu/All-Net/english/cardpage/operate/bypass/intro.htm)
- [http://pedsccm.wustl.edu/All-Net/english/ekgpage/ekg-4a.htm](http://pedsccm.wustl.edu/All-Net/english/ekgpage/ekg-4a.htm)
- [http://www.google.co.uk/search?hl=en&q=Cardioplegia+solutions+in+paediatric+cardiopulmonary+bypass+&meta=](http://www.google.co.uk/search?hl=en&q=Cardioplegia+solutions+in+paediatric+cardiopulmonary+bypass+&meta=)
Information for Year 2 ITU Training (advanced):

Year 2 ITU curriculum

- DORV
- Aortopulmonary Window

Curriculum Notes for Year 2:

DOUBLE-OUTLET RIGHT VENTRICLE

Double-outlet Right Ventricle describes a ventriculo-arterial alignment. It is a heterogeneous group of lesions with variable physiology and surgical management.

Characteristic feature - The great vessels are side by side and parallel. This is a result of the subaortic conus which causes the aorta to lie more anterior and to be aligned with the right ventricle. As a result, the two valves are at the same level and
there is **aortic-mitral valve discontinuity**. DORV is always associated with a VSD. At one end of the spectrum, the DORV with subaortic VSD shares many features in common with TOF. At the other end of spectrum, DORV with subpulmonary VSD shares many features in common with TGA. The categorisation of DORV is based on the relationship of VSD to the semilunar valves; however it is important to recognise the most important morphologic factor that determines different types of DORV is not the variation of VSD but the varying orientation of the great vessels based on the degree of rotation of the conotruncus.

**There are 4 types of DORV**

1. Sub-Aortic VSD, with or without pulmonary stenosis
2. Subpulmonary VSD, with or without subaortic stenosis and/or arch obstruction
3. Doubly committed VSD
4. Remote VSD

**Sub-Aortic VSD, with or without pulmonary stenosis**
The VSD directs left ventricular outflow to the aorta, the degree of PS determines symptoms
- If moderate to severe PS – cyanosis
- If little or no PS – congestive heart failure
- Or balanced – enough PS to ‘protect the pulmonary arteries from overcirculation and pulmonary hypertension

**Surgery**
- Closure of VSD to baffle the left ventricular outflow to aorta, with relief of pulmonary outflow tract obstruction as determined by the anatomy.
- The right ventricular infundibulum is opened and the baffle is placed around the anterolateral and inferior rims of the VSD thereby closing VSD and creating left ventricular to aortic continuity.
- Depending on the status of development of the subpulmonary conus, the infundibular incision can be patched or closed primarily.

**Post-operative Issues**
Usual complications of TOF
Exclude subaortic obstruction caused by either a small VSD or the VSD baffle itself.

**Sub-pulmonary VSD, with or without sub-aortic stenosis and/or arch obstruction**
Physiologically similar to d-TGA
The VSD directs LV outflow back to the pulmonary artery, whereas the systemic venous return recirculates to the aorta. Because of large VSD and abundant intercirculatory mixing, cyanosis may be absent and symptoms of CCF may predominate. Sub-aortic stenosis and/or arch obstruction typically complicate this anatomic subtype.

**Surgery**
Closure of VSD to baffle LV outflow to the pulmonary root
Arterial Switch operation with or without arch augmentation
The posterior translocation of the native aorta to the neo-pulmonary root can cause proximal arch obstruction.

**Post-operative Issues**
Sequelaes of arterial Switch with VSD closure  
Subaortic obstruction  
Subpulmonic obstruction  
Arch obstruction

**DORV with Doubly committed VSD or Remote VSD**
Complex repairs needed. Frequently impossible to baffle LV outflow to either vessel without significant intracardiac obstruction; in these patients staged management to eventual Fontan operation should be considered.

**AORTOPULMONARY WINDOW**

**Definition**: A vascular communication between the ascending aorta and the main pulmonary artery. This communication can be variable in size and location (either close to the semilunar valves or to the right pulmonary artery).

**Associated lesions**: Can be associated with VSD, coarctation and interruption of the arch. Rarely aortic origin of the RPA can be associated. Also, one or both coronary arteries can arise anomalously from either the aorticopulmonary window or the pulmonary artery.

Patients usually have symptoms and signs of congestive heart failure after the fall in pulmonary vascular resistance in the first weeks of life. Because of the large size of the defect, pulmonary hypertension can be irreversible as early as 1 year of life.

The defect is closed with a patch with care to avoid the coronary arteries.

**Postoperative Management**
Usually uneventful. Rarely postoperative pulmonary hypertension. Please see management as described in the section on AVSD.

**Websites.**
http://www.emedicine.com/ped/topic2813.htm