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Tricuspid Atresia

16.1 Morphology

Tricuspid atresia (TA) is characterized by the absence or agenesis of the tricuspid valve. A dimple or a localized fibrous thickening in the floor of the right atrium at the expected site of the tricuspid valve occurs in 90% of cases (muscular *type*). Other types of TA *(membranous, valvular, Ebstein, and AV* (*atrioventricular) canal*) account for the rest 10%.

RA (right atrium) is enlarged and hypertrophied. An interatrial communication (most commonly PFO (patent foramen ovale) or some times the secundum or primum ASD (atrial septal defect)) allows obligatory shunting of the blood to the left atrium. The left ventricle is morphologically normal but enlarged and hypertrophied. VSD (ventricular septal defect) is present commonly (perimembranous or malaligned type between the limbs of the septal band), and it can be large. RV (right ventricle) is composed of only an infundibulum and a trabecular portion, which communicates with the left ventricle (through VSD). The inflow region of RV is absent. RV is hypoplastic and small, but the RV size varies with anatomic type of TA. It is large with a large VSD or transposition of the great arteries, and is very small with pulmonary atresia and normally related great arteries.

TA is classified into four main types by the relationship of great arteries and each type is further subgrouped into three, by the degree of pulmonary outflow tract obstruction that is present in most cases.

Type I: Normally related great arteries.

Type II: D-Transposition of the great arteries.

Type III: Great artery positional abnormalities other than D-transposition (e.g., L-transposition and anatomically corrected transposition (SLL).

Type IV - Persistent truncus arteriosus.

The subgroups in all types are:

Subgroup a: Pulmonary atresia.

Subgroup b: Pulmonary stenosis or hypoplasia.

Subgroup c: Normal pulmonary arteries without stenosis (see Figure 16.1).

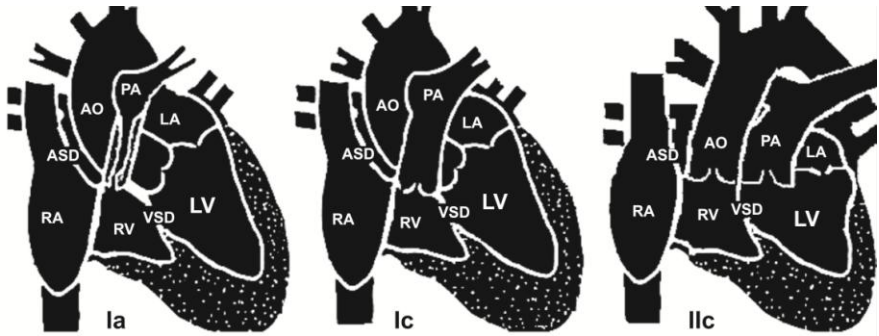

Figure 16.1 *Diagrammatic representation of types of tricuspid atresia. Note the absence of tricuspid valve. The presence of ASD allows obligatory right to left shunting of systemic venous blood. VSD which is usually of a large size allows shunting of blood from left to the right ventricle. Type I a: tricuspid atresia with pulmonary atresia and normally related great arteries. Type Ic: tricuspid atresia without pulmonary stenosis and normally related great arteries. Type IIc: tricuspid atresia without pulmonary stenosis and transposed (D) great arteries. AO= aorta, PA=pulmonary artery, RA=right atrium, LA=left atrium, RV=right ventricle, LV=left ventricle, ASD=atrial septal defect, VSD=ventricular septal defect.*

16.2 Pathophysiology

The obligatory shunting of systemic venous blood across PFO or ASD into the left atrium causes admixture of systemic venous and pulmonary venous returns. This blood then passes onto the left ventricle across the mitral valve. This flow pattern occurs in all types but type III (with L transposition or anatomically corrected transposition (SLL).

In type I, shunting through a VSD permits pulmonary blood flow. In the absence of a VSD, the pulmonary blood flow is derived through patent ductus arteriosus or aortopulmonary collateral vessels. The systemic blood flow is derived directly from the left ventricle. In type II, the pulmonary blood flow is directly from the left ventricle. The systemic blood flow is from the left ventricle through VSD and the right ventricle.

16.2.1 Pulmonary Outflow Tract

The degree of pulmonary outflow tract obstruction and patency of the ductus arteriosus determine the quantity of pulmonary blood flow.

In type Ia, the pulmonary flow is markedly decreased and presents with signs of severe cyanosis, hypoxemia, and acidosis.

In type Ic (with a large and nonrestrictive VSD without pulmonary stenosis) and type II, the pulmonary blood flow is increased and present with signs of heart failure later in infancy. The LV (left ventricle) ejects the entire systemic and pulmonary outputs so volume overloading is present, and pulmonary flow is inversely proportional to the pulmonary-to-systemic vascular resistance ratio.

16.2.2 Obstruction of Interatrial Communication

As the entire systemic venous blood must egress through an interatrial communication, it is not unlikely that an interatrial obstruction may develop. Obstruction is probable if > 5 mm Hg pressure gradient develops between atria with prominent “a waves” in the jugular venous pulse, presystolic hepatic pulsations, and hepatomegaly. If a restrictive interatrial communication develops, causing systemic venous congestion, tanscatheter or surgical atrial septostomy may be needed as an immediate palliation.

16.2.3 Ventricular Septal Defect

Patency of a VSD is essential to maintain intracardiac shunting and for patient survival. VSD closure in type I results in progressive cyanosis, increasing polycythemia, and diminution or disappearance of the murmur of VSD. Intermittent functional closure of the VSD results in cyanotic spells in TA. Closure of the VSD in type II (transposition) produces subaortic (i.e., systemic outflow) obstruction. Partial or complete spontaneous closures have been reported.

16.2.4 Systemic Arterial Oxygen Saturation

It is decreased in all TA's because of an obligatory admixture of systemic and pulmonary venous blood in the left atrium. The degree of arterial desaturation depends on the amount of pulmonary blood flow. A Qp: Qs ratio of 1.5 to 2.5 may result in adequate in oxygen saturation, but the higher pulmonary flow (high Qp: Qs ratios) produces left ventricular volume overloading without an increase in oxygen saturation.

16.3 Clinical Presentation

Cyanosis: It occurs in the first few days of life in infants with severe decrease in pulmonary blood flow (PBF). Tachypnea and acidosis are present if the PBF is markedly decreased. Most patients have a type Ib defect. In type Ic, cyanosis appears early as the ductus begins to close.

Dyspnea & fatigue: These symptoms, suggestive of congestive heart failure, difficulty in feeding, perspiration, failure to thrive, and recurrent respiratory tract infection develop within a few to several weeks in infants with increased PBF. Most patients have type IIc with a VSD, some have type Ic. Aortic coarctation may be present in some of the type II defects with early onset of CHF (congestive heart failure).

16.4 Medical Management

16.4.1 Prostaglandin E_1 Infusion

It should be initiated to treat severe hypoxemia due to decreased pulmonary blood flow.

16.4.2 Mechanical Ventilation

It may be needed if severe hypoxemia and acidosis complicates cyanosis.

16.4.3 Digitalis and Diuretics

These drugs are used to treat severe congestive heart failure due to increased PBF until an operative intervention can be undertaken to restrict the pulmonary blood flow.

16.5 Surgical Management

16.5.1 Palliative Correction

Cyanosis & hypoxemia with decreased pulmonary blood flow:

a) Systemic-pulmonary artery shunt (Blalock-Taussig shunt)/or

b) Cavopulmonary anastomosis (Glenn shunt).

The bidirectional Glenn is often indicated for recurrent symptoms of cyanosis or increased volume load after a prior palliation if the patient is < 18 months old. If subaortic obstruction develops due to closure of the VSD in type II (transposition), aortopulmonary anastomosis (Damus-Kay Stansel procedure) and bidirectional cavopulmonary anastomosis are indicated.

Left ventricular volume load& CHF with increased pulmonary blood flow:

Pulmonary artery banding may supplement medical treatment of congestive heart failure. It is often required in type II c and rarely, in type Ic.

Note: About 50% of the patients survive to teen years by performing above palliative procedures to control PBF, but the risk for developing complications of the disease (i.e., paradoxical emboli, stroke, brain abscess, polycythemia, progressive cardiac dilatation, ventricular dysfunction, mitral valve insufficiency, and arrhythmias) is much increased.

16.5.2 Definitive Surgical Correction

Fontan procedure & Indications:

The shunt failure (recurrent cyanosis, progressive polycythemia, decreasing exercise tolerance) or increasing pulmonary obstruction is an indication for reevaluation and consideration for a second palliative procedure or the Fontan procedure.

Fontan Operation:

1. The direct right atrial-to-pulmonary artery connection with or without a conduit excluding the right ventricle.
2. Lateral tunnel Fontan if the patient had a previous bidirectional cavo-pulmonary anastomosis or
3. The extracardiac cavopulmonary anastomosis by a synthetic graft excluding the RV.

Fontan is done if criteria are met and the following parameters may generally ensure a good outcome:

a) Patient should be > 18 months or 4 years older and in sinus rhythm, without pulmonary artery distortions from previous palliative procedures.

b) The pulmonary artery-to-aorta diameter ratio should be greater than 0.75.

c) Mean pulmonary artery pressure should be low (i.e., < 15 mm Hg).

16.6 Postoperative Management

16.6.1 Palliative Procedures

Adequate pulmonary blood flow is of paramount importance and monitor for signs of hypoxemia (i.e., discrease PBF) or pulmonary plethora (i.e., increase PBF).

Pulmonary vascular resistance/ systemic vascular resistance (PVR / SVR):

Balance the ratio of PVR/SVR to 1:1 to maintain adequate systemic perfusion and pulmonary blood flow. May perform echocardiographic assessments to determine the patency and adequacy of the shunt if the ratio of PVR/SVR is < 1.

Signs of hypoxemia in the form of rising hemoglobin levels must be monitored on the long term for early detection of a shunt failure.

Early detection of CHF by clinical evaluation may suggest increased pulmonary blood flow with increased ratio of PVR/SVR.

16.6.2 Fontan Operation: (See Also Univentricular Heart, Chapter 17)

Maintain adequate blood flow through the Fontan circuit to ensure the adequate LV output.

During the immediate postoperative period (first 12 to 24 hours), maintain adequate preload (CVP of 15 mm HG) until the adequate perfusion is established with warm extremities and hemodynamic stability. During the later postoperative period, fluid volume load is restricted and diuresis is encouraged.

Pulmonary vascular resistance: All efforts are made to maintain pulmonary vascular resistance as low as possible using supplemental oxygen and pulmonary vasodilators.

Promptly address the causes that elevate PVR during the immediate postoperative period such as, hypoxia, hypercapnia, pain, agitation, etc.

LV Contractility:

Maintain adequate LV function by inotropic support.

Cardiac out is assessed by indirect means (base deficit, temperature of toes and extremities, urine output) or directly.

Heart rhythm: Maintain sinus rhythm to optimize the cardiac output (see Section I Chapter 4).

Volume load: Monitor for signs of hepatic congestion which can occur secondary to the operative procedure per se and the subsequent volume overload.

If the signs of volume load appear or CVP > 15 mm Hg is noted, initiate infusions of NTG (nitroglycerin) and/or pulmonary vasodilators such as PGE_1 or nitric oxide. Early and aggressive treatment with diuretics is indicated.

Digitalis and Diuretics:

These drugs control the congestive heart failure that is often present.

Digitalis maintains sinus rhythm and improves cardiac contractility.

Diuretics are often administered to treat and prevent pulmonary edema.

Invasive monitors:

Arterial, central venous, and LA (left atrial) catheters.

Vasoactive Drug infusions:

Dopamine or dobutamine, epinephrine, milrinone, and nitroprusside (see Section I Chapters 4 & 16).

Cardiac Rhythm Abnormalities:

Sinus node dysfunction:

Extensive atrial suture lines and alteration of the normal intra-atrial pressures can lead to a loss of sinus rhythm and results in sinus node dysfunction. It is common following Fontan completion. Sinus node dysfunction commonly presents as sinus arrhythmia, severe tachycardia, and loss of a normal atrioventricular synchrony (see Section I Chapter 4).

Temporary overdrive pacing:

It is needed as hemodynamic compromise is common in the Fontan patient subsequent to loss of normal atrioventricular synchrony or in severe tachycardia.

Permanent pacemaker: It is occasionally required.

Postoperative Bleeding:

Excessive bleeding is very rare.

Anticoagulation:

Slow venous blood flow through long segments of prosthetic material make the Fontan patients susceptible to thrombus formation in the early postoperative period.

Coumadin is started in Fontan patients once intracardiac lines have been removed.

Following a cavopulmonary shunt, the anticoagulation is less rigorous and might include only aspirin or dipyridamole.

Pleural effusion: Patients often develop effusion which should be monitored carefully, and is removed in order to maximize oxygenation and decrease pulmonary vascular resistance.

Persistent pleural drainage following a bidirectional cavopulmonary shunt is rare, but is common following a Fontan completion so bilateral chest tubes may be routinely placed in both pleural spaces during the Fontan completion.

Chemical Pleurodesis:

If pleural drainage continues more than 10 days or even a week, the chemical pleurodesis may be required to control protracted pleural drainage (see Section I Chapter 12).

Discharge:

In uncomplicated Fontan patient or a bidirectional cavopulmonary shunt, the discharge medications should include coumadin or antiplatelet agent, digoxin, lasix, and captopril.