Pulmonary venous anomalies



Total anomalous pulmonary venous returns (TAPVR)

- All pulmonary venous blood flow returns anomalously to the systemic veins or directly to the right atrium
- Prevalence estimated at 1 in 10,000
- Acutely cyanotic infant in shock
- One of the true surgical emergencies across the entire spectrum of congenital heart surgery.



TAPVR

- Biventriclular heart
- Single ventricle
- Heterotaxy syndrome



TAPVR = total anomalous pulmonary venous return; VSD = ventriculoseptal defect PAPVR = partial anomalous pulmonary venous return; COA = coarctation of the aorta TOF = tetralogy of Fallot; DROV = double outlet right ventricle

Embryology

EMBRYOLOGY



Anatomic subtype

- Type 1 : supracardiac (43-50%)
- Type 2 : Cardiac (18-20%)
- Type 3 : Infracardiac type (20-27%)⁻
- Type 4 : Mixed (10-12%)
- Non-opstructed vs. Obstructed



Type 1 : Supracardiac type

- Vertical vein most often drains to LIV
- Course between LPA and left main bronch
- May present obstructed (around 50%)





TAPVR(to innominate, obstructive type)







Type 2 : Cardiac type

- Typically to the coronary sinus
- · Less likely to be obstructed
- Can present later



Type 3: Infracardiac type

- Descending vein to portal vein, IVC, hepatic vein, or ductus venosus
- Nearly all obstructed \rightarrow present at birth





Type 4 : mixed type







TAPVR (mixed type)



TAPVR (Rt. Isomerism)





Diagnosis

- Echocardiography
- Cardiac angiography
- CT
- Cardiac MRI

Key feature of echo



Small appearing Left heart

Pure $R \rightarrow L$ shunt at PFO

Key feature of ech.o



PV veins to confluence

Type 2 – to the CS





"Pulmonary hypertension"







The importance of the Atrial communication



All preload to the LV is supplied by right to left shunting across the atrial communication

Cardiac catheterization – stenting vertical vein



CT angiogram





- The superior approach
- Lateral technique
- "In situ" technique
- Sutureless repair
- Primary sutureless Repair
- Lateral approach
- Surgery for pulmonary venous obstruction after repair of TAPVC

Superior approach







Transverse sinus approach





Lateral approach



Lateral approach



Lateral approach



"In situ" technique



"In situ" technique





COMPREHENSIVE SURGICAL MANAGEMENT OF CONGENITAL HEART DISEASE second edition



RICHARD A. JONAS ILLUSTRATED BY REBEKAH DODSON





Sutureless repair



Surgery for post-repair pulmonary vein stenosis

a)









Surgery for post-repair pulmonary vein stenosis







Primary sutureless repair



Primary sutureless repair





Cardiac type repair

Cardiac type repair

Cardiac type repair

- TAPVR repair
- Separate patch closure

Postop. management

- Consideration of muscularized pulmonary arteries (obstructive TAPVC)
- Minimization of pulmonary resistance
 - Appropriate ventilator care (PCO2 level)
 - Oxygen, NO gas
 - Low dose isoproterenol (pulmonary vasodilatory effect)
 - Sedation
- Careful observation for pulmonary hypertensive crisis

Prognosis

- Early mortality : 8-20%
- Risk factors for early mortality
 - Preoperative pulmonary venous obstruction
 - Single ventricle anatomy
 - Chromosomal anomaly
 - Small pulmonary confluence
 - Diffuse pulmonary vein
- Pulmonary vein stenosis : 10-20%
 - Presence of preoperative obstruction
 - Endocardial sclerosis (recurrent obstruction)

Partial anomalous pulmonary venous return

- M/C associated with sinus venosus ASD
- Usually, RUL draining to SVC
- Rare anomaly in right pulmonary veins
 - Single vertical trunk descends in a curve to enter the IVC (Scimitar syndrome)
- No symptoms and signs when Qp/Qs is less than 1.5

PAPVR

Partial anomalous pulmonary venous return

RIGHTS RESERVED.

PAPVR

Scimitar syndrome

Scimitar Syndrome.

PAPVR repair

PAPVR repair

PAPVR repair

Warden procedure

Herbert E. Warden 1984

Repair of scimitar syndrome

Possible complications

- SVC obstruction
- Pulmonary venous stenosis
- Sinus node dysfunction
- Sick sinus syndrome

Cor Triatriatum

- 1. Communication with LA
 - No other connection
 - Other connection
 - 1. To RA
 - 2. TAPVC
- 2. No communication with LA
 - Connection to RA
 - TAPVC

Cor Triatriatum

