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Superior repair: A useful approach for some anatomic variants of total anomalous pulmonary venous connection

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ABSTRACT

Background: Total anomalous pulmonary venous connection (TAPVC) occurs when all the four pulmonary veins drain to the right atrium or to tributaries of the systemic veins. There have been various published techniques for the repair but none has been agreed on for the different anatomical variants that may be encountered during surgery. **Patients and Methods:** Between January 2005 and June 2010 the data of 6 of 18 patients who had surgical repair using the superior approach were retrospectively reviewed.Three patients had long narrow venous stalk connecting the coronary sinus to venous confluence; two had the right pulmonary veins draining to superior vena cava (SVC) and left pulmonary veins to left lateral wall of SVC and one had an obstruction at entrance of Pulmonary Vein into venous confluence. **Results:** Five patients initially had the superior approach while one had transatrial with unroofing of the coronary sinus. Two had a concomitant Wardens procedure. The mean aortic cross clamping was 87.5 (60-125) min, the mean cardiopulmonary bypass time should have min as unit of 127.8 (100-180), the mean Intensive Care Unit (ICU) stay of 2.5 (2-4) days and the mean hospital stay 8.2 (7-9) days. One patient died at early post-operation from low cardiac outputand five had an uneventful post-operative course and had remained stable until date. **Conclusion:** In our experience, the superior approach was an effective alternative approach for some anatomic variants of TAPVC that may be unexpectedly encountered during operation and also useful surgical approach for older children.

Key words: Long venous stalk, pulmonary venous obstruction, superior approach, trans-atrial approach, warden's procedure

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INTRODUCTION

Total anomalous pulmonary venous connection (TAPVC) is a condition in which all the pulmonary veins drain into the right atrium either directly or indirectly rather than the left atrium. It makes up about 1-1.5% of the children with congenital heart diseases.[1,2] Various techniques of repair have been advocated over the years.[3-6] The use of high-frequency transducers combined with Doppler colour flow mapping were found to be reliable tool in the diagnosis of TAPVC, and surgical repairs were decided on the basis of the findings from the echocardiography.[7] Despite the reliability of the diagnostic tools, anatomic variants can be accidentally diagnosed intra-operatively and may pose a challenge to the unwary Surgeon during surgery. Although surgical outcomes of TAPVC have improved over the years as a result of accumulated knowledge of the disease and advanced perioperative management strategy; these variants can still alter the outcome of surgical treatment especially, amongst the older patients.[7,8]

PATIENTS AND METHODS

Between January 2005 and June 2010 the data of 6 of 18 patients who had surgical repair using thee superior approach and were discovered intra-operatively to have some specific anatomic variants of TAPVC were analysed. One of the patients had unroofing of the coronary sinus at the first operation and was readmitted with pulmonary venous obstruction (PVO)3 months later and subsequently had repair using the Superior approach. At surgery, it was observed that the obstruction was at the entrance of pulmonary veins into venous confluence (the pre-operative echocardiography did not show that). Three patients were discovered, intra-operatively, while using the superior approach to have a long narrow venous stalk connecting the coronary sinus to the venous confluence. The remaining two patients had the right pulmonary veins connected to superior vena cava (SVC) and left veins to a venous

confluence connected to left lateral wall of SVC; they also had Warden's procedure in addition because of the right pulmonary veins connected to the SVC. The Warden's procedure involved achieving anastomosis between the transected proximal portion of the SVC and the amputated apex of right atrial appendage with the use of absorbable sutures and subsequent over sewing the transected distal portion of SVC above the opening of partial anomalous pulmonary venous connection (PAPVC). A synthetic patch or autologous pericardium is used to suture the margin of the sinus venosus defect, effectively baffling the SVC orifice to direct blood flow from the anomalous pulmonary veins, through the conduit, through the sinus venosus defect into the left atrium.[9]

Surgical technique

The superior repair was through a median sternotomy under cardiopulmonary bypass established using aortic and bicaval cannulation (in nominate vein). The vertical vein was identified and looped and the SVC mobilized with the azygos vein divided between ligatures for increased visibility. The main pulmonary, right pulmonary and left pulmonary arteries were mobilized and the patient ductusarteriosus divided. The pulmonary vein confluence was identified and the anatomy of the venous drainage was assessed with the aorta and SVC retracted apart to gain adequate space. The SVC or Aorta may also be divided if space is a constraint; though in our series there was no occasion for that in any of the patients. Myocardial protection was ensured by doses of cold blood cardioplegia delivered to the aortic root. The ascending aorta and the pulmonary artery were retracted laterally, and the roof of the left atrium exposed with a tie placed on the left atrial appendage to facilitate exposure. The posterior pericardium just superior to the dome of the left atrium was incised, and the pulmonary venous confluence was identified with a longitudinal incision made along the entire length of the confluence to create a wide opening. A matching incision was made on the superior aspect of the roof of the left atrium, placing gentle traction leftward on the left atrial appendage. The anastomosis between common pulmonary vein and roof of left atrium (LA) using 6/0 or 7/0prolene or maxon was performed. The suture line was started at the leftward extent and carried along the superior edge of the atriotomy and the inferior edge of the venous confluence and completed by joining the two remaining edges. The atrial septal defect was closed through the opening in the LA roof or through the Right Atrium. The patient was slowly weaned off bypass and the vertical vein ligated in supracardiac type of TAPVC. Heparin reversed with protamine and routine chest closure carried out. Warden's procedure was performed for partial anomalous pulmonary venous connection and it involved redirection of the right pulmonary veins into the LA by transecting the right SVC and anastomosing it to right atrial appendage and closing the SVC and ASD thereby committing the right pulmonary veins to the left atrium.[5,7] The approach allowed us to perform the two procedures at the same time.

RESULTS

The age ranged from 1 month to 17 years; the male to female ratio was 5:1 with the mean aortic cross clampingtime was 87.5 (60-125) min, the mean Cardiopulmonary bypass time was 127.8 (100-180), the mean ICU stay was 2.5 (2-4) days and the mean hospital stay was 8.2 (7-9) days.One of the patients died immediately after surgery from low cardiac output and the other five of the patients had uneventful post-operative recovery and have remained asymptomatic as at the last follow-up time at 1 year period [Table 1].

Table 1: The bio-data and clinical outcome

Age	Sex	Clinical features	Diagnosis	Anatomic variant	AXC min	CPB min	Outcome
10 month	M	dyspnea, poor feeding, and cyanosis	cardiac	Long stalk	85	122	Alive
17 year	M	SOB, stunted growth, easy fatiguability, cyanosis	Supracardiac	RPV	95	125	Alive
1 month	M	dyspnea, poor feeding, and cyaosis	cardiac	OPV	125	180	Dead**
3 year	F	dyspnea, failure to thrive, and cyanosis	cardiac	Long stalk	70	110	Alive
10 month	M	dyspnea, poor feeding, and poor growth	Supracardiac	RPV	90	130	Alive
2.9 year	M	dyspnea, failure to thrive, cyanosis	cardiac	Long stalk	60	100	Alive

OPV: Obstruction at entrance of pulmonary vein into confluence, RPV: Right pulmonary vein draining into SVC and the pulmonary vein draining in the left side SVC, AXC: Aortic cross clamp time, CPB: Cardiopulmonary bypass time, SOB: Shortness of breath,**Died of low cardiac output, SVC: Superior vena cava

DISCUSSION

There are different anatomic variants of TAPVC and their surgical options.[5,8-11] Occasionally, the diagnoses of these variants can easily be missed during the pre-operative evaluation of the patient using echocardiography, or patients may present late in adulthood.[7,8] Consequently, the managements of these patients are determined largely by the experience of the surgeon and the surgical options adopted.

When the transatrial approach was used in our institution, it was attended by some problems such as difficult access, intraoperative bleeding and anastomic site narrowing post-operatively with consequent pulmonary congestion. However, with the adoption of the superior approach, it was possibly to ensure good exposure of the roof of the left atrium as well as the common pulmonary vein (the venous confluence) and create better anastomosis even in patients with some of these anatomic variants. Furthermore, it was useful for re-do surgeries for the same problem as it offeredthe advantage of not excessively displacing the heart with also easy of applicability in older children without having tomake incision across cristal terminalis at the right atriumas would have beenwith the transatrial approach[5-8,12]

In one of the cases [Table 1], the patients hadlong venous stalk connecting the Venous confluence to the coronary sinus, the stalk could have easily be obstructedat any point along the entire length and approaching this variant by unroofing of the coronary sinus may still have left the stalk susceptible to obstruction and, possibly, not identified; but with the superior approach it was possible for the inspection of the confluence at surgery with identification of the entire length of the stalk.

The two patients who had the right pulmonary veins emptying into the right side of cavoatrial junction andthe left pulmonary veins forming a confluence thatalso emptied into the left side of cavoatrial junction with an atrial septal defect; with the superior approach, the venous confluence was opened and anastomosed to leftatriumthereby ensuring adequate connection without narrowing as would have resulted from the tension that might have ensued from the combination ofthe transatrial approach with the Warden's procedure.

PVO with occasional pulmonary hypertension may predominate after correction of obstructed type of TAPVC post-operatively, and also no anatomic arrangements are spared and the anatomic and pathologic changes.[12-19] So, using the superior approach ensured the creation of wider anastomosis devoid of tension thereby averting the effect of these anatomical and pathologic changes after surgery.

CONCLUSION

In our experience, the superior approach was an effective alternative approach for some anatomic variants of TAPVC that may be unexpectedly encountered during operation and also useful surgical approach for older children.

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