




Technical Details of Rechannelling of Obstructive Coronary Sinus Type of Totally Anomalous Pulmonary Venous Connection Using Malm's Coronary Sinus Cutback Technique and Left Atrial Augmentation: A Video Presentation

Ujjwal K. Chowdhury¹  Niwin George¹ Sushamagayatri B.¹ Balaji Chandhirasekar¹ Shikha Goja¹ Nagasai Manjusha¹ Niraj Nirmal Pandey² Poonam Malhotra Kapoor³

¹Department of Cardiothoracic and Vascular Surgery, Cardiothoracic Sciences Centre, All India Institute of Medical Sciences, New Delhi, India

²Department of Cardiac Radiology, Cardiothoracic Sciences Centre, All India Institute of Medical Sciences, New Delhi, India

³Department of Cardiac Anaesthesia, Cardiothoracic Sciences Centre, All India Institute of Medical Sciences, New Delhi, India

Address for correspondence Ujjwal K. Chowdhury, MCh, Diplomate NB, Department of Cardiothoracic and Vascular Surgery, All India Institute of Medical Sciences, New Delhi 110029, India (e-mail: ujjwalchowdhury@gmail.com).

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Abstract

Keywords

- ▶ left atrial augmentation
- ▶ Malm's coronary sinus cutback technique
- ▶ rechannelling of obstructive coronary sinus
- ▶ totally anomalous pulmonary venous connection

Obstructive coronary sinus totally anomalous pulmonary venous connection although rare, is associated with a high mortality. A 2-month-old female child diagnosed with obstructive coronary sinus type of totally anomalous pulmonary venous connection and severe pulmonary arterial hypertension underwent complete unroofing of the coronary sinus by the “coronary sinus cutback technique” of Malm with left atrial augmentation and atrial septal fenestration.

Introduction

Totally anomalous pulmonary venous connection accounts for 1.3 to 3% of cases of chronic heart disease.^{1,2} Infants born with totally anomalous pulmonary venous connection have a

generally unfavorable prognosis, with only 50% surviving beyond 3 months and 20% surviving the first year of life.^{3–5}

Hazelrig et al analyzed data from 183 autopsied cases of surgically untreated totally anomalous pulmonary venous connection reported in the literature.⁶ Median survival was

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2 months with the shortest survival being 1 day and the longest 49 years. Ninety percentage of deaths occurred in the first year of life. Obstruction of the pulmonary venous pathway reduced the median survival from 2.5 months in the nonobstructed group to 3 weeks in the obstructed group. Patients with supracardiac and cardiac connections had a similar natural history with median survival of 2.5 and 3 months, whereas those with infracardiac connection had median survival of 3 weeks.¹⁻⁵

Obstruction of pulmonary veins draining to the coronary sinus is poorly recognized and generally considered extremely rare.⁷⁻⁹ This impression has largely developed from autopsy series containing few surgical patients and almost no patients who died late after surgery.¹⁻³ In carefully analyzed surgical series, however, the incidence of obstructed coronary sinus drainage has been between 22 and 36%.¹⁰⁻¹⁴

Cross-sectional echocardiography and color Doppler studies are versatile tools with 100% sensitivity and 85% specificity in detecting the presence of obstruction.⁸ Obstruction is manifested by the demonstration of a narrowed segment in the pulmonary venous pathway and the presence of a nonphasic Doppler spectrum or turbulent flow in the pulmonary venous confluence, vertical vein, or coronary sinus.^{8,11,13,14} Cardiac catheterization probably should be reserved for instances when pulmonary hypertension is detected by echocardiography and the level of obstruction cannot be established.¹⁴⁻¹⁶

The long-term outcome of obstructive cardiac totally anomalous pulmonary venous connection is favorable when the obstruction is relieved at the initial operation and the prognosis becomes grim when new onset obstruction develops in the postoperative period.^{12,17}

Relevant studies in the published literature on the optimal surgical technique to be utilized in obstructive coronary sinus type of totally anomalous pulmonary venous connection are lacking. Some investigators have demonstrated the development of obstruction at the site of "fenestration" following Van Praagh's fenestration technique.¹¹ Because complete unroofing following the traditional coronary sinus "cut back technique" allows improved visualization of the internal junction of the pulmonary veins with coronary sinus, this technique as proposed by James Malm may be preferable to the fenestration method of Van Praagh or intra-atrial rerouting technique of Yamagishi et al.¹⁷⁻²⁰

We report herein a 2-month-old female child diagnosed with obstructive coronary sinus type of totally anomalous pulmonary venous connection and severe pulmonary arterial hypertension undergoing complete unroofing of the coronary sinus by the "coronary sinus cutback technique" of Malm with left atrial augmentation and atrial septal fenestration. Postoperatively, the child was in normal sinus rhythm and recovery was uneventful.

Surgical Techniques

Following median sternotomy, the thymus was subtotally excised taking care not to expose the brachiocephalic vein.

The pericardium was opened in the midline in between stay sutures using scissors and not cautery to avoid inadvertent cautery-induced ventricular fibrillation.

The operation was performed with moderately hypothermic cardiopulmonary bypass through angled venous cannulas into superior and inferior caval veins and aortic cannulation. Cold hyperkalemic blood cardioplegia and topical hypothermia are used for myocardial preservation.

The persistent ductus arteriosus is ligated using No.2 ductus silk suture pulling down the superior surface of the pulmonary artery at the commencement of cardiopulmonary bypass as described by Dwight McGoon. The pump flow is temporarily lowered at the time of ligation of the ductus arteriosus.

Following aortic cross-clamp and administration of cardioplegia, the right atrium was opened longitudinally parallel to the atrioventricular groove. The atriotomy incision was intentionally placed quite forward and extended from the right atrial appendage (not from the superior vena cava) to avoid injury to the sinoatrial node, Bachmann's bundle, and the course of the posterior intermodal tract in the lateral atrial wall.

The coronary sinus was large, ellipsoidal, and approximately 2 cm long and 1.5 cm wide. A right-angle clamp was inserted into the markedly enlarged coronary sinus. The conjoined anterior wall of the coronary sinus and posterior wall of the left atrium was pushed with the tip of the clamp so that the conjoined wall was displaced through the patent foramen ovale.

The septum between the coronary sinus and patent foramen ovale was excised, as described by James Malm, thus creating a wide communication between the coronary sinus and left atrium. Extreme precaution need to be taken during incision of the posterior left atrial wall not to incise the pulmonary venous orifices to avoid postoperative pulmonary venous obstruction. To avoid damage to the atrioventricular node, a longitudinal incision along its caudal edge is avoided. The string of superior limbic band between the coronary sinus and patent foramen ovale is not excised to avoid injury to the middle internodal tract.

The opening of all four pulmonary veins was identified. The size of the opening of the unroofed coronary sinus with the atrial septal defect was measured in both vertical and horizontal direction.

A redundant Dacron polyester patch (Bard Savage filamentous knitted polyester fabric, Bard Peripheral Vascular Inc., Tempe, Arizona, United States), little larger than the size of the defect was used to divert all pulmonary venous blood, together with the coronary sinus to the left atrium. Anteriorly, 5-0 polypropylene suture (Johnson and Johnson Ltd., Ethicon, LLC, San Lorenzo, United States) runs through the floor of the coronary sinus away from its anterior rim so as to avoid injury to the tail of the atrioventricular node. The right side of the patch was deviated to the body of the right atrium away from the margin of the atrial septal defect. This maneuver enhances the capacity of the left atrium.

A 2-mm calibrated atrial septal fenestration was performed for decompression of the right-sided chambers in the event of pulmonary hypertensive crisis. The right atrium was closed in

two layers using 5–0 polypropylene. The aortic cross-clamp was released, thus restoring blood flow to the myocardium. The chest is primarily closed in layers (–Video 1).

Video 1

Showing Coronary sinus cutback technique in TAPVC
 Online content including video sequences viewable at:
<https://www.thieme-connect.com/products/ejournals/html/10.1055/s-0042-1759806>.

Short-Term and Long-Term Results

Pulmonary hypertension was treated with hyperventilation, sedation, phenoxybenzamine, sildenafil, and inhaled nitric oxide at 10 to 15 PPM, in varying combination for 48 hours. Postoperatively, the patient was hemodynamically stable on dopamine (7.5 µg/kg/min), dobutamine (7.5 µg/kg/min), and milrinone at a dose of 50 µg/kg intravenous bolus followed by 0.375 to 0.75 µg/kg/min. Postoperative echocardiography demonstrated an entirely satisfactory repair, with a large anastomosis, with no gradient between the pulmonary venous confluence and left atrium and nonturbulent biphasic pulmonary venous flow at < 1.2 m/s.

The child was extubated on second postoperative day with stable hemodynamics. At 26 months' follow-up, the child was asymptomatic, showed no clinical evidence of cardiac failure, with Ross's clinical score of 2, and without antifailure cardiac medications. Echocardiography revealed normal biventricular function and absence of flow through the atrial septal fenestration.

Conclusion

The traditional coronary sinus cutback technique allows complete unroofing of the coronary sinus in obstructive cardiac totally anomalous pulmonary venous connection without bradyarrhythmias. This method is safe, expedient, provides an optimal exposure, and may be the technique of choice in cases of obstructive coronary sinus type of totally anomalous pulmonary venous connection.

Conflict of Interest

None declared.

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