



GLOBAL JOURNAL OF MEDICAL RESEARCH: I
SURGERIES AND CARDIOVASCULAR SYSTEM
Volume 19 Issue 1 Version 1.0 Year 2019
Type: Double Blind Peer Reviewed International Research Journal
Publisher: Global Journals
Online ISSN: 2249-4618 & Print ISSN: 0975-5888

A New Palliative Surgical Technique for High Risk Total Anomalous Pulmonary Venous Connection (Sarmast-Takriti Shunt)

By Dr. Hossein Sarmast & Dr. Ahmad Takriti

Damascus University

Abstract- Objectives: Total Anomalous Pulmonary Venous Connection (TAPVC) is a rare heterogeneous condition. That accounting for 1.5-3% of congenital heart diseases. It is characterized by failure of the Pulmonary Venous Confluence (PVC) to be directly connected to the left atrium in combination with a persistent splanchnic connection to the systemic venous circulation. The most critical status occurs when it is accompanied by pulmonary venous obstruction. Managing of this situation is very difficult and in fact, pulmonary venous obstruction is usually lethal. The real aim of this study is offering a new palliative surgical technique (Sarmast – Takriti Shunt) in order to alleviate the patient's signs and symptoms until becomes read for the main surgical correction.

Methods: The study included a 4-day old, low birth weight boy who suffered from Critical Obstructive Total Anomalous Pulmonary Venous Connection. The decision was made to perform the new palliative technique using Gore-Tex (ePTFE). Anastomosis was established without Cardiopulmonary Bypass (CPB) between Pulmonary Venous Confluence (PVC) and the left atrial appendage. Therefore the Sarmast – Takriti Shunt (STS) was taken place.

Keywords: total pulmonary venous connection, low birth weight, sarmast-takriti shunt (STS), pulmonary venous obstruction, pulmonary venous confluence palliative surgical technique.

GJMR-I Classification: NLMC Code: WB 310



ANWPALLIATIVE SURGICAL TECHNIQUE FOR HIGH RISK TOTAL ANOMALOUS PULMONARY VENOUS CONNECTIONS ARMAS TAKRITI SHUNT

Strictly as per the compliance and regulations of:



RESEARCH | DIVERSITY | ETHICS

© 2019. Dr. Hossein Sarmast & Dr. Ahmad Takriti. This is a research/review paper, distributed under the terms of the Creative Commons Attribution-Noncommercial 3.0 Unported License (<http://creativecommons.org/licenses/by-nc/3.0/>), permitting all non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

A New Palliative Surgical Technique for High Risk Total Anomalous Pulmonary Venous Connection (Sarmast-Takriti Shunt)

Dr. Hossein Sarmast^α & Dr. Ahmad Takriti^σ

Abstract- Objectives: Total Anomalous Pulmonary Venous Connection (TAPVC) is a rare heterogeneous condition. That accounting for 1.5-3% of congenital heart diseases. It is characterized by failure of the Pulmonary Venous Confluence (PVC) to be directly connected to the left atrium in combination with a persistent splanchnic connection to the systemic venous circulation. The most critical status occurs when it is accompanied by pulmonary venous obstruction. Managing of this situation is very difficult and in fact, pulmonary venous obstruction is usually lethal. The real aim of this study is offering a new palliative surgical technique (Sarmast – Takriti Shunt) in order to alleviate the patient's signs and symptoms until becomes ready for the main surgical correction.

Methods: The study included a 4-day old, low birth weight boy who suffered from Critical Obstructive Total Anomalous Pulmonary Venous Connection. The decision was made to perform the new palliative technique using Gore-Tex (ePTFE). Anastomosis was established without Cardiopulmonary Bypass (CPB) between Pulmonary Venous Confluence (PVC) and the left atrial appendage. Therefore the Sarmast – Takriti Shunt (STS) was taken place.

Results and Conclusions: After completion of the procedure, the pressure gradient across the venous confluence and the Left innominate vein became zero. Cyanosis, agitation and feeding Problem subsided. Three days later, when he was discharged, arterial oxygen saturation had reached as high as 91%. After seven months we performed the main correction.

Keywords: total pulmonary venous connection, low birth weight, sarmast-takriti shunt (STS), pulmonary venous obstruction, pulmonary venous confluence palliative surgical technique.

I. INTRODUCTION

Total anomalous pulmonary venous connection is a rare heterogeneous anomaly, accounts for 1.5–3% of congenital heart diseases (1). It is characterized by abnormal return of whole pulmonary venous blood flow to the right atrium or systemic venous tributaries due to its persistent splanchnic connection (2). A concomitant right to left shunt, commonly via an Interatrial communication, is required for survival after birth. Darling classified it in four

Author α: MD, Resident of Cardiovascular Surgery in Cardiac Surgery Hospital of Damascus University, Mouasat Square, Omar ben Abdulaziz Street, Damascus, Syria. e-mail: abcmoghim@gmail.com

Author σ: MD, Full Professor and Chief of Cardiac Surgery Department in Cardiac Surgery Hospital of Damascus University. e-mail: takritiahmad@gmail.com

categories: Supra-cardiac 45%, cardiac 25%, Infra-cardiac 25% and mixed type 5-10% (3).

At one end of the spectrum, there are completely unobstructed circulation, these neonates present with a large left to right shunt manifestations. At the other end there are severe PVO. Neonates born with TAPVC have poor prognosis with approximately 80% mortality in the first year of life. Both obstructed and non-obstructed types of TAPV pose an absolute indication for surgical repair (4). In PVO type without intervention the median survival is two months, with the shortest survival being 1 day. Despite greatly improved neonatal care and surgical techniques over the last decade, TAPVC operation is still associated with high hospital mortality, up to 20% (5-6).

II. CASE PRESENTATION

A 4 – day old, low birth weight boy (w = 1950 gr) was presented to our department with discrete but increasing cyanosis, tachypnea, respiratory distress, hepatomegaly, hypoxia (SaO₂=70%), gasping, poor feeding and severe metabolic acidosis. The prenatal course was uneventful and he was born by normal vaginal delivery on gestational age=38.5 w.

The patient didn't carry any congenital heart disease (CHD) history in his genetically close relatives (first, second and third degree). Immediate and brief work up was carried out. Chest X Ray (CXR) showed normal heart size with ground glass appearance in all the lung fields (fig.1. a). Color Doppler and 2D- echocardiography revealed the total anomalous pulmonary venous connection (TAPVC – supra cardiac type), accompanied by significant gradient between the drainage point of vertical vein to the left brachiocephalic vein and the pulmonary veins with flow acceleration > 3.0 m/sec (pulmonary venous obstruction). It was also uncovered presence of the ASD secundum, as the natural last resort for being alive. The vertical vein was noted to be compressed as it coursed posterior the left pulmonary artery and anterior the left main bronchus (fig. 1.b). According to the aforementioned findings, the boy had almost met most of incremental risk factors leading to mortality after conventional operations.

Therefore the decision was made to a new palliative surgical procedure for the first time. In order to preoperative medical stabilizing we administered 100% O₂ with the aim of promoting respiratory alkalosis as well as nitric oxide as a pulmonary vascular dilator, since the patient had severe metabolic acidosis besides Pulmonary hypertension (PHT). Under general anesthesia, median sternotomy and partial thymectomy were carried out. The pericardium was opened in vertical fashion then prudent purse-string sutures as standby were placed on ascending aorta and right atrial appendage (without using CPB). After intravenous heparinization (100 U/kg), at first some dissections were done from left lateral side between heart and pulmonary venous confluence then the dome of the left atrium was exposed. The posterior pericardium just superior the dome of LA was incised and PVC was appeared (fig. 2). Using a side – biting clamp on the PVC, a longitudinal incision was made. The proximal head of a Gore-Tex (ePTFE) with appropriate size (diameter= 6 mm) that had been prepared and beveled, was anastomosed to PVC using continuous 6-0 polypropylene suture. Under topical cooling of heart and using a side-biting clamp on left atrial appendage (LAA), the distal end of Gore-Tex was anastomosed to LAA. After deairing with heparinized saline as routine, the clamp was removed. The Sarmast-Takriti Shunt (STS) between PVC and LA was established (fig. 3). Immediately after completion of the procedure, cyanosis began to decrease. We performed the main operation 7-months later with excellent outcome when he had already sustained satisfactory weight (w= 7030 gr), as follows: After the establishment of CPB, the shunt was removed. To reduce the risk of residual obstruction of PVC due to pocket-like contraction our team preferred modified septosuperior approach (komarakshi technique). A direct anastomosis between PVC and L. A, ligation of the VV and closure of ASD with autopericardial patch were achieved in one stage repair.

III. RESULTS AND DISCUSSION

Evaluation of pressures before intervention in the operation room and after correction are illustrated in the (table.). Immediately after completion of surgery (STS), the pressure of PVC decreased to the point where its pressure gradient became zero. Blood oxygenation improved up to 84% (preoperative SaO₂ was 70% on 100% oxygen) and cyanosis, agitation, feeding Problem subsided. Three days later, when he was discharged, arterial oxygen saturation had reached as high as 91%. Despite good advances in treating of TAPVC in recent decades, this severe malformation in its various anatomical forms remains a challengeable entity during early infancy. Significant obstruction to pulmonary venous drainage results in pulmonary edema in the presence of a normal size and shape of the heart and cardiogenic shock which is rapidly lethal if

untreated. Almost all reports have declared that perioperative high mortality associates with PVO, low weight (W<2.5-3 kg), early age (A<2m), severe preoperative acidosis, long time of Aortic Cross Clamp (ACC) and cardiac arrest. The second frontier in the treatment of TAPVC is represented by postoperative PVO. In such a difficult situations, if patients survive from operation, most of them will require multiple postoperative surgical interventions due to recurrent PVO with an increasingly poor outcome each representation (7). Medical efforts are minimally effective in managing the ensuing hemodynamic and metabolic problems so their use is limited to provide some short lived conservative therapy until definitive surgical treatment is carried out. PVO is usually lethal, even with reoperation and extensive attempts at revision or repair (8). This lack of success has led to alternative treatments such as balloon dilatation and stenting. The Rashkind Operation or Balloon Atrial Septostomy (BAS) has been used with some success to decompress the pulmonary venous pressure and improve C/O in the restricted ASD, but these don't appear to provide additional benefit. Moreover several reports have proposed the use of percutaneous angioplasty and stenting of the obstructed vein to palliate shock and improve preoperative metabolic state. Research showed during the median cross - sectional follow up of 3.1 years estimated mortality was 38+/- 8% at 1 year and 50+/- 8% at 5-years after stent implantation.

Necessity for reintervention (owing to occlusion of stent), was 58+/-7% at 1-year. In 1996 sutureless repair technique was described, using in situ autologous pericardium for recurrent pulmonary vein stenosis following main TAPVC surgery (9). Subsequent reports emphasize the utility of this technique in selected patients as main procedure. Despite interest in the sutureless technique, there is little firm evidence that it provides a benefit over conventional techniques used a retrospective analysis to compare the outcomes of death and restenosis after conventional and sutureless techniques. By multivariable analysis, there was no statistically significant difference between the conventional and sutureless techniques. We encountered with a patient, who had almost encompassed all critical risk factors that were sufficient to make the operative prognosis very poor. We believed that Sarmast-Takriti Shunt (STS) would ensure adequate postoperative hemodynamics for symptomatic neonate and prompt left cardiac side rehabilitation. The STS with confined heparin (100U/kg), was carried out without using CPB with an intention to reduce the morbidity associated with extra corporeal circulation. Eliminating CPB reduced the cost of the procedure substantially and saved the patient from its inherent complications. (10) After procedure the enough time was prepared on behalf of the heart to compensate its chambers especially the right ventricle and left atrium and ensure endurable state

for the main surgery. Although our experience was limited to STS in supracardiac type, we are optimistic and hopeful to its feasibility and usefulness in other types of TAPVC. Now, we are so satisfied owing to be able to help such a complicated neonate.

ACKNOWLEDGEMENTS

The author wish to thank: Professor Zahra Sepehrmanesh, Dr. Soroush Sarmast & Dr. Behina Sarmast for their assistance in this study. Also Dr. Kevin Brady & Dr. Fariba Brady from the USA state of New Jersey; For their editorial assistance.

Funding statement

As we are from lower income country, the processing charge has been waived.

Conflict of interest statement

None declared.

REFERENCES RÉFÉRENCES REFERENCIAS

1. St Louis J D, Harvey B A, Menk J S, Raghuvver G, O'Brien J E Jr, Bryant R 3rd, Kochilas L. Repair of "simple" total anomalous pulmonary venous connection: a review from the Pediatric Cardiac Care nConsortium. *Ann Thorac Surg.* 2012; 94: 133–137. doi: 10.1016/j.athorac sur.2012.03.006.
2. Kouchoukos N T, Blackstone E H, Hanley F L, Kirklin JK. *Kirklin/BarratBoyes Cardiac Surgery.* 4th edn. Philadelphia: Elsevier Saunders, 2013, 1182–207.
3. Darling R C, Rothney W B, Craig J M. Total pulmonary venous drainage into the right side of the heart; report of 17 autopsied cases not associated with other major cardiovascular anomalies. *Lab Invest* 1957; 6: 44–64.
4. Shi G, Zhu Z, Chen J, Ou Y, Hong H, Nie Z et al. Total anomalous pulmonary venous connection: the current management strategies in a pediatric cohort of 768 patients. *Circulation* 2017; 135: 48–58.
5. Duff J P, Joffe A R, Vatanpour S, Moddemann D M, Robertson C M, Alton G et al. Neurocognitive outcomes at kindergarten entry after surgical repair of total anomalous pulmonary venous connection in early infancy. *Pediatr Cardiol* 2015; 36: 350–7.
6. Cui H J, Chen X X, Ma L, Xia Y S, Yang S C, Zou M H, et al. Surgical treatment of total anomalous pulmonary venous connection under 6 months of age. *Chin J Surg.* 2016; 54(4): 276 –80.
7. Kato H, Fu Y Y, Zhu J, Wang L, Aafaqi S, Rahkonen O, Slorach C, Traister A, Leung C H, Chiasson D, Mertens L, Benson L, Weisel R D, Hinz B, Maynes J T, Coles J G, Caldarone C A. Pulmonary vein stenosis and the pathophysiology of "upstream" pulmonary veins. *J Thorac Cardiovasc Surg.* 2014; 148: 245–253. doi: 10.1016/j. jtcvs.2013.08.046.
8. Masuda M, Okumura M, Doki Y, Endo S, Hirata Y, Kobayashi J et al. Thoracic and cardiovascular surgery in Japan during 2014: annual report by The Japanese Association for Thoracic Surgery. *Gen Thorac Cardiovasc Surg* 2016; 64: 665–97.
9. Gao X M, Nie Z Q, Ou Y Q, He B C, Yuan H Y, Qu Y J, et al. Comparison between two surgical techniques to repair total anomalous pulmonary venous connection using propensity score analysis. *Sun Yat-Sen Univ.* 2017; 38(1): 143 –50.
10. Sakamoto T. Current status of brain protection during surgery for congenital cardiac defect. *Gen Thorac Cardiovasc Surg* 2016; 64: 72–81.

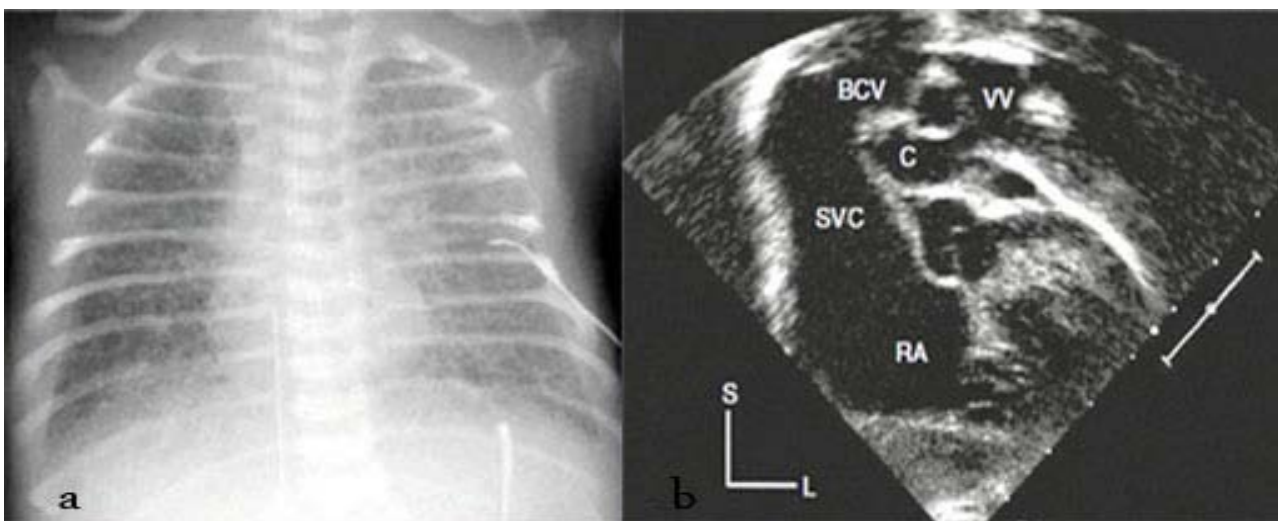


Figure 1: 4-days old male with total anomalous pulmonary venous connection accompanied by pulmonary venous obstruction: a- CXR: Note mild cardiac enlargement and evidence of pulmonary venous hypertension ("ground glass" appearance). b- 2D-Echocardiography shows compressed vertical vein between left pulmonary artery and left main bronchus.

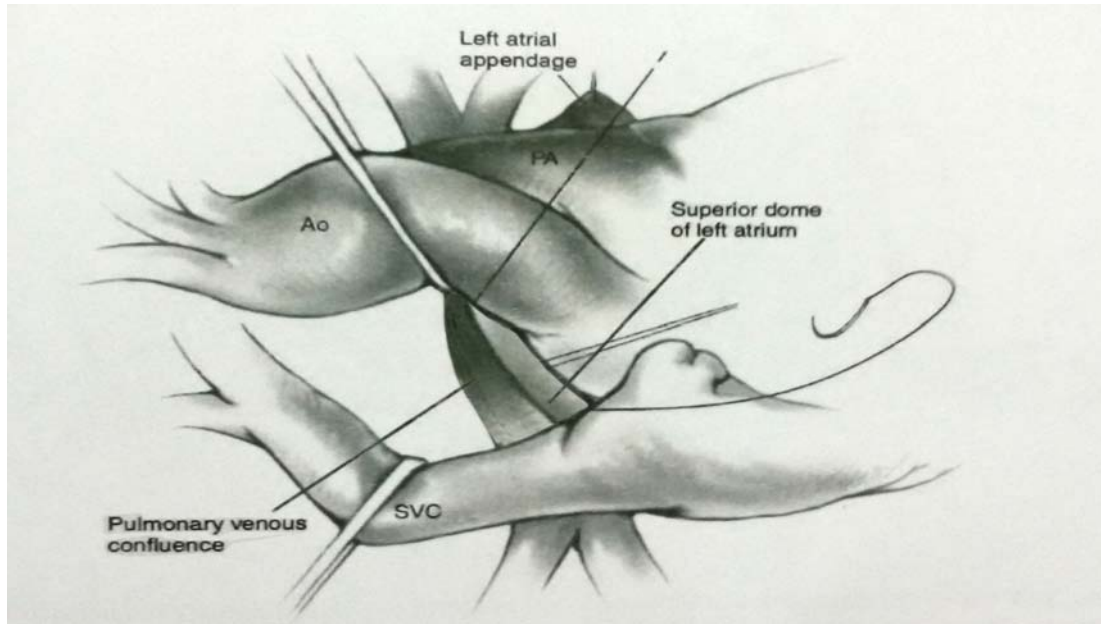


Figure 2: Schematic illustration of superior approach in 4-days old male with total anomalous pulmonary venous connection.

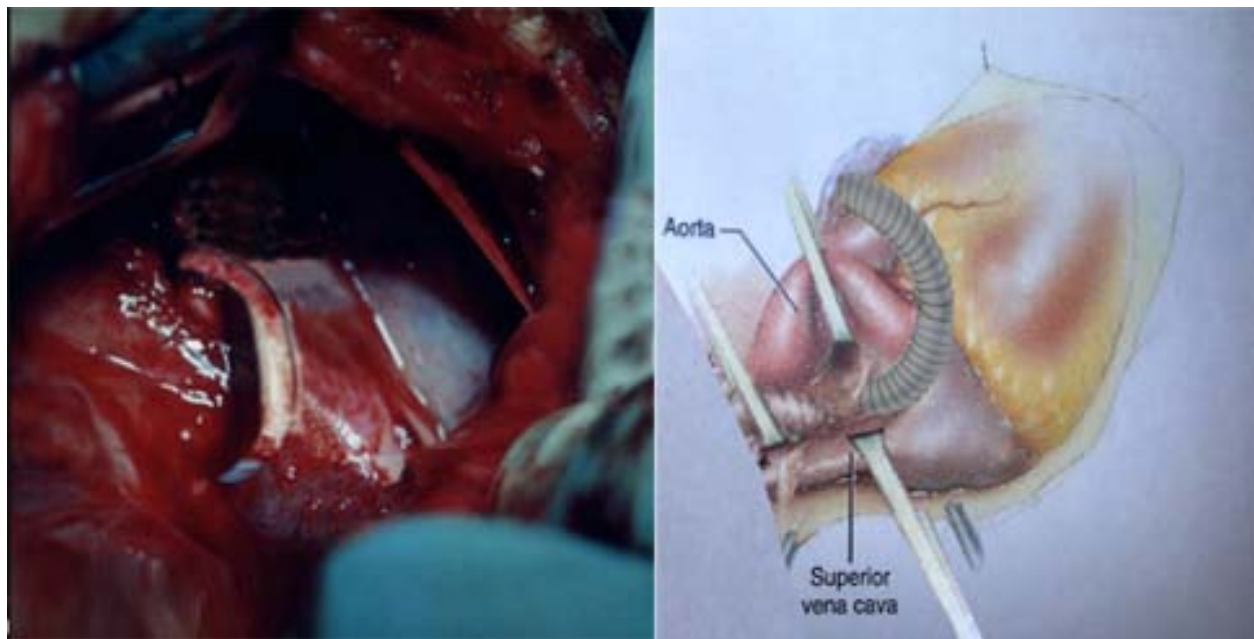


Figure 3: Schematic and operational photo illustrations of sarmast - takriti shunt (STS) in 4-days old male with total anomalous pulmonary venous connection.

Table 1: Preoperation and postoperative cardiac pressures of 4-days old male with total anomalous pulmonary venous connection accompanied by pulmonary venous obstruction.

	PVC Mean Pressure	Left Brachiocephalic Vein	Left Atrium	Right Atrium	Right Ventricle	Pulmonary Artery
Preoperative pressures (mmHg)	29	9	8	9	61/13	59/31
Postoperative pressures (mmHg)	8	8	9	8	32/10	28/15