

Interactive CardioVascular and Thoracic Surgery

**Total anomalous pulmonary venous drainage with major aorto-pulmonary
collaterals – a diagnostic dilemma**

Sanjay Kumar, Junaid Ansari, Mike E.C. Blackburn and Nihal Weerasena
Interact CardioVasc Thorac Surg 2005;4:484-485; originally published online Jul 18,
2005;
DOI: 10.1510/icvts.2005.108670

The online version of this article, along with updated information and services, is
located on the World Wide Web at:

<http://icvts.ctsnetjournals.org/cgi/content/full/4/5/484>

Interactive Cardiovascular and Thoracic Surgery is the official journal of the European Association
for Cardio-thoracic Surgery (EACTS) and the European Society for Cardiovascular Surgery
(ESCVS). Copyright © 2005 by European Association for Cardio-thoracic Surgery. Print ISSN:
1569-9293.

Case report - Congenital Total anomalous pulmonary venous drainage with major aorto- pulmonary collaterals – a diagnostic dilemma

Sanjay Kumar^{a,*}, Junaid Ansari^a, Mike E.C. Blackburn^b, Nihal Weerasena^a

^aDepartment of Cardiothoracic Surgery, Yorkshire Heart Centre, Leeds General Infirmary, Great George Street, Leeds, LS1 3EX, UK

^bDepartment of Pediatric Cardiology, Yorkshire Heart Centre, Leeds General Infirmary, Great George Street, Leeds, LS1 3EX, UK

Received 6 March 2005; received in revised form 29 May 2005; accepted 6 July 2005

Abstract

A term female newborn presented with right atrial isomerism and infra-diaphragmatic total anomalous pulmonary venous drainage associated with complex cardiac anatomy. The repair was performed utilizing circulatory arrest in deep hypothermia. However, in the post-operative period the patient could not be weaned off mechanical ventilation and underwent cardiac catheterisation. This revealed major aortopulmonary collaterals, which were occluded with coils. We review the literature with specific focus on the occurrence of this unusual combination and its implication in the management during the postoperative period.

© 2005 Published by European Association for Cardio-Thoracic Surgery. All rights reserved.

Keywords: Major aorto-pulmonary collaterals; Right atrial isomerism; Total anomalous pulmonary venous drainage; Infracardiac total anomalous pulmonary venous drainage; Heterotaxy syndrome

1. Introduction

Right atrial isomerism with total anomalous pulmonary venous drainage (TAPVD) is an uncommon cardiac anomaly [1,2]. TAPVD associated with complex cardiac anatomy has a markedly less favourable prognosis when compared to isolated TAPVD [2]. The association of major aortopulmonary collaterals (MAPCAs) in right atrial isomerism with infra-cardiac TAPVD is unknown. We review the literature specifically with its implication on the management in the postoperative period.

2. Case summary

A female Caucasian neonate, weighing 2880 g, was born at term with an antenatal diagnosis of complex congenital heart disease (CCHD). She developed cyanosis shortly after birth with oxygen saturations of 30% and was started on prostaglandins (PGE₂). She was ventilated and transferred to the regional pediatric cardiac unit.

On arrival, echocardiography confirmed CCHD with dextrocardia, right atrial isomerism, complete atrioventricular septal defect (AVSD), infundibular and valvar pulmonic stenosis (PS), double outlet right ventricle (DORV) with left ventricle dominance, and infra-diaphragmatic TAPVD (Darling type III). An urgent repair of the infracardiac TAPVD was performed on the third day of life.

A median sternotomy was performed. Cardio-pulmonary bypass was initiated with aorto-right atrial cannulation and

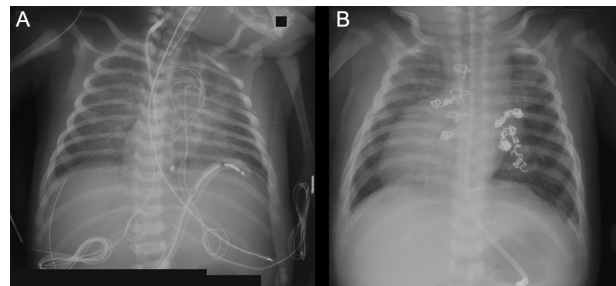


Fig. 1. A. Post repair pre-catheterisation chest X-ray showing plethoric lung fields. B. Post coiling chest X-ray with reduced plethora.

the repair was performed under deep hypothermic circulatory arrest (DHCA) at 18 °C. The common atrium was opened and a wide anastomosis was performed between the atrium and the common venous chamber. The vertical vein was ligated. The CP Bypass and DHCA time were 120 and 35 min, respectively. Postoperatively, her hemodynamics stabilized with minimal inotropic support. Further recovery was delayed by chylothorax and coagulase negative staphylococcal sepsis.

A major concern was her ventilator dependency two weeks post TAPVD repair despite maintaining oxygen saturation above 94%. Chest X-ray showed pulmonary plethora (Fig. 1A) and echocardiograms confirmed good repair. In view of the unexplained pulmonary plethora and inability to wean her off mechanical ventilation, a cardiac catheterisation and angiography was performed. It confirmed echocardiogram findings. Pulmonary arteries were hypo-

*Corresponding author. Tel.: +44 7980631248; fax: +44 1133928092.

E-mail address: sanjaykr33@hotmail.com (S. Kumar).

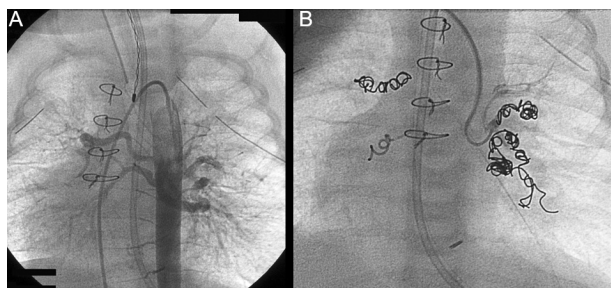


Fig. 2. A. Angiogram showing four MAPCAs. B. Angiogram after the coil occlusion of all four MAPCAs.

plastic and both lungs were additionally supplied by MAPCAs (Fig. 2A). She underwent percutaneous transcatheter coil occlusion of four MAPCAs using a total of 26 embolism coils (Fig. 2B). The right upper lobe was solely supplied by collaterals from the right subclavian artery and was therefore not embolised. Post embolisation chest X-ray showed reduced pulmonary blood flow (Fig. 1B). She was successfully weaned off the ventilator and was subsequently discharged back to her local hospital. On follow-up at 12 months of age, she is doing remarkably well.

3. Discussion

Heterotaxy syndromes are associated with complex congenital heart disease with high morbidity and mortality. TAPVD is associated with 87% of these patients [1]. Infra-cardiac TAPVD constitutes about 33% of total TAPVD [2]. Despite improvement in survival for simple cases, management of TAPVD with single-ventricle hearts or other associated cardiac lesions remains challenging [2]. Over the decade operative mortality for simple cases decreased from 26% to 8%, however, mortality for complex cases remained constant at 52% [2]. Multivariable analysis identified only univentricular hearts and associated cardiac lesions as predictors of operative mortality [3]. A more aggressive approach is suggested by early recognition and assessment of these critically ill infants, followed by early surgery [3].

Pulsed Doppler ultrasound has been used to characterise pulmonary venous flow patterns in the foetus with TAPVD and associated complex congenital cardiac lesions [4]. Echocardiography is a reliable tool for the assessment of these cyanotic neonates. In neonates with TAPVD, the drainage sites and flow profiles of the pulmonary veins can be exactly determined by Doppler echocardiography preoperatively, which makes cardiac catheterisation unnecessary [5]. Cardiac catheterisation is reserved for cases where the diagnosis is a mixed TAPVD [6]. Helical computed tomographic angiography with differential colour imaging technique and three-dimensional reconstruction is an alternative to cardiac catheterisation [7]. It provides precise spatial information of complicated vascular anomalies and clearly demonstrates pulmonary venous obstruction in an infant with TAPVD [7].

There is no mention of successful correction of a combination of infra-cardiac TAPVD with MAPCAs in the literature so far. This unique association with a dramatic impact on the post-repair pathophysiology seems to be due to a

developmental insult early in the embryonic formation of the supporting vasculature of the lung. A structural study of pulmonary circulation and of the heart in TAPVD in early infancy using quantitative morphometric techniques led to some interesting observations. In infra-diaphragmatic TAPVD, obstruction to pulmonary venous return develops soon after birth and prevents a large increase in pulmonary blood flow, and thus neither the pulmonary arteries nor the right ventricle become dilated [8]. The lung tissue from TAPVD patients with pulmonary venous obstruction has demonstrated hypoplasia of small pulmonary arteries, which elucidate the mechanism underlying the poor outcome in these patients [9]. In addition, association of PS, like in the present case, must have led to decreased pulmonary blood flow in the foetal life and hence development and persistence of MAPCAs.

4. Conclusion

We conclude that the need for prolonged ventilation following repair of an unobstructed TAPVD with the evidence of increased pulmonary flow should strongly raise the suspicion of MAPCAs. There is a need to rule out preoperatively the possible association of MAPCAs when PS and TAPVD coexist. Cardiac catheterisation and angiography play a vital role in the diagnosis and treatment of such cases.

Acknowledgements

We thank Mr S Powell and Ms Ezenee Kolbaba, Department of Medical Illustration, LGI, Leeds Teaching Hospitals, Leeds, United Kingdom for their help with illustration.

References

- [1] Hashmi A, Abu-Sulaiman R, McCrindle BW, Smallhorn JF, Williams WG, Freedom RM. Management and outcomes of right atrial isomerism: a 26-year experience. *J Am Coll Cardiol* 1998;31(5):1120–1126.
- [2] Caldarone CA, Najm HK, Kadletz M, Smallhorn JF, Freedom RM, Williams WG, Coles JG. Surgical management of total anomalous pulmonary venous drainage: impact of coexisting cardiac anomalies. *Ann Thorac Surg* 1998;66(5):1521–1526.
- [3] Sinzobahamvya N, Arenz C, Brecher AM, Blaschczok HC, Urban AE. Early and long-term results for correction of total anomalous pulmonary venous drainage (TAPVD) in neonates and infants. *Eur J Cardiothorac Surg* 1996;10(6):433–438.
- [4] Feller Printz B, Allan LD. Abnormal pulmonary venous return diagnosed prenatally by pulsed Doppler flow imaging. *Ultrasound Obstet Gynecol* 1997;9(5):347–349.
- [5] Saxena A, Reddy SC, Kothari SS, Juneja R, Venugopal P, Shrivastava S. Mixed variety of total anomalous pulmonary venous connection: diagnosis by 2D echocardiography and Doppler colour flow imaging. *Indian Heart J* 1999;51(1):65–68.
- [6] Imoto Y, Kado H, Asou T, Shiokawa Y, Tominaga R, Yasui H. Mixed type of total anomalous pulmonary venous connection. *Ann Thorac Surg* 1998;66(4):1394–1397.
- [7] Shiraiishi I, Yamagishi M, Iwasaki N, Toyama K, Hamaoka K. Helical computed tomographic angiography in obstructed total anomalous pulmonary venous drainage. *Ann Thorac Surg* 2001;71(5):1690–1692.
- [8] Haworth SG, Reid L. Structural study of pulmonary circulation and of heart in total anomalous pulmonary venous return in early infancy. *Br Heart J* 1977;39(1):80–92.
- [9] Maeda K, Yamaki S, Yokota M, Murakami A, Takamoto S. Hypoplasia of the small pulmonary arteries in total anomalous pulmonary venous connection with obstructed pulmonary venous drainage. *J Thorac Cardiovasc Surg* 2004;127(2):448–456.

Total anomalous pulmonary venous drainage with major aorto-pulmonary collaterals – a diagnostic dilemma

Sanjay Kumar, Junaid Ansari, Mike E.C. Blackburn and Nihal Weerasena
Interact CardioVasc Thorac Surg 2005;4:484-485; originally published online Jul 18, 2005;
DOI: 10.1510/icvts.2005.108670

This information is current as of February 9, 2010

| | |
|---|---|
| Updated Information & Services | including high-resolution figures, can be found at: http://icvts.ctsnetjournals.org/cgi/content/full/4/5/484 |
| References | This article cites 9 articles, 7 of which you can access for free at: http://icvts.ctsnetjournals.org/cgi/content/full/4/5/484#BIBL |
| Subspecialty Collections | This article, along with others on similar topics, appears in the following collection(s): Lung - other http://icvts.ctsnetjournals.org/cgi/collection/lung_other Cardiac - physiology http://icvts.ctsnetjournals.org/cgi/collection/cardiac_physiology Cardiac - other http://icvts.ctsnetjournals.org/cgi/collection/cardiac_other Congenital - cyanotic http://icvts.ctsnetjournals.org/cgi/collection/congenital_cyanotic |
| Permissions & Licensing | Requests to reproducing this article in parts (figures, tables) or in its entirety should be submitted to: icvts@ejcts.ch |
| Reprints | For information about ordering reprints, please email: icvts@ejcts.ch |

Interactive CardioVascular and Thoracic Surgery