

8-2013

EReReply. Re: Balancing the pulmonary circulation in tricuspid atresia with an aortopulmonary window

Pranava Sinha

Children's National Medical Center, Washington, DC

Syed Murfad Peer

Children's National Medical Center, Washington, DC

Mary T. Donofrio

George Washington University

Follow this and additional works at: http://hsrc.himmelfarb.gwu.edu/smhs_peds_facpubs



Part of the [Pediatrics Commons](#)

Recommended Citation

Sinha, P., Peer, S. M., Donofrio, M. T., & Gaur, L. (2013). EReply. re: Balancing the pulmonary circulation in tricuspid atresia with an aortopulmonary window. *Interactive Cardiovascular and Thoracic Surgery*, 17(2), 443.

This Journal Article is brought to you for free and open access by the Pediatrics at Health Sciences Research Commons. It has been accepted for inclusion in Pediatrics Faculty Publications by an authorized administrator of Health Sciences Research Commons. For more information, please contact hsrc@gwu.edu.

cardiopulmonary bypass on the pulmonary vasculature and the developing neonatal brain. Closure of the AP window was a requisite measure to reduce the symptoms of heart failure in this patient.

Increasing restriction at the level of a VSD in patients with tricuspid atresia may be due to dynamic or mechanical obstruction or as a consequence of alteration in ventricular geometry following volume unloading surgery, as described in congenital heart defects with the double inlet left ventricle or tricuspid atresia with the VSD and transposition of the great arteries [5]. We believe that the AP window ligation, though providing an adequate initial first step for single ventricle surgical palliation, may have decreased the volume load to the left ventricle in our patient, thus adding to the evolving subpulmonary stenosis, and may have contributed to the need for the Blalock–Taussig shunt. However, alternative approaches such as the previously described fenestrated repair of the AP window [1] would leave multiple sources of pulmonary blood flow with their unpredictability; or complete separation of the pulmonary circulation with the addition of systemic-to-pulmonary shunt would leave the child shunt-dependent. Additionally, both approaches would require the use of cardiopulmonary bypass and add to the complexity of the procedure.

Our surgical approach to AP window closure was initially successful; however, the progression of disease resulted in the need for an additional surgical palliative procedure. In summary, our experience highlights the complexities in the management of neonates with such complex single ventricle disease.

Conflict of interest: none declared.

REFERENCES

- [1] Geva T, Ott DA, Ludomirsky A, Argyle SJ, O’Laughlin MP. Tricuspid atresia associated with aortopulmonary window: controlling pulmonary blood flow with a fenestrated patch. *Am Heart J* 1992;123:260–2.
- [2] Sittiwangkul R, Azakie A, Van Arsdell GS, Williams WG, McCrindle BW. Outcomes of tricuspid atresia in the Fontan era. *Ann Thorac Surg* 2004; 77:889–94.
- [3] Kutsche LM, Van Meirop LHS. Anatomy and pathogenesis of aorticopulmonary septal defect. *Am J Cardiol* 1987;59:443–7.
- [4] Patent ductus arteriosus, aortopulmonary window, sinus of Valsalva fistula, aortoventricular tunnel. In: Jonas RA (ed). *Comprehensive Surgical Management of Congenital Heart Disease*. 1st edn. London, UK: Arnold, 2004, 187–206.
- [5] Donofrio MT, Jacobs ML, Norwood WI, Rychik J. Early changes in ventricular septal defect size and ventricular geometry in the single left ventricle after volume-unloading surgery. *J Am Coll Cardiol* 1995;26:1008–15.

eComment. Balancing the pulmonary circulation in tricuspid atresia with an aortopulmonary window

Author: Sandeep Sainathan

Albert Einstein College of Medicine/Bronx-Lebanon Hospital, New York, NY, USA
doi:10.1093/icvts/ivt247

© The Author 2013. Published by Oxford University Press on behalf of the European Association for Cardio-Thoracic Surgery. All rights reserved.

It was interesting to read the article by Peer *et al.* on "Tricuspid atresia with aortopulmonary window: challenges in achieving a balanced circulation" [1]. The authors are commended in dealing with a difficult situation of tricuspid atresia with an aortopulmonary window, as there are no established treatment guidelines, given the extreme rarity of this condition. Tricuspid atresia with pulmonary over-circulation is known with a 30% incidence. The present case represents such a situation. While

every attempt was made to keep the procedure extracardiac to balance pulmonary circulation, simple ligation of the aortopulmonary window was feasible in this case due to a favourable anatomy. However, simple suturing, besides not definitely treating the condition, carries a risk associated with the distortion of the left coronary ostia or the right pulmonary artery. The child had three consecutive procedures to address the whole problem. An alternative strategy would have been to leave the aortopulmonary window intact with restricting the pulmonary over-circulation with bilateral branch pulmonary artery banding. A similar approach with an univentricular physiology has been described for hypoplastic left heart syndrome [2]. Even if the ventricular septal defect would have become restrictive, as it happened in this case, pulmonary circulation via the aorto-pulmonary window would have been maintained. This may have potentially saved an extraoperative procedure of creating a systemic to pulmonary shunt. The child could then be palliated finally with a definitive repair of the aorto-pulmonary window and tracked along a modified Fontan pathway.

Conflict of interest: none declared

References

- [1] Peer SM, Donofrio MT, Gaur L, Sinha P. Tricuspid atresia with aortopulmonary window: challenges in achieving a balanced circulation *Interact CardioVasc Thorac Surg* 2013;17:441–3.
- [2] Hosein RB, Mehta C, Stickley J, Mcguirk SP, Jones TJ, Brawn WJ, Barron DJ. Creation of aorto-pulmonary window with pulmonary artery band is not good palliation for hypoplastic left heart syndrome. *Eur J Cardiothorac Surg* 2007;32:745–50.

eReply. Re: Balancing the pulmonary circulation in tricuspid atresia with an aortopulmonary window

Authors: Pranava Sinha, Syed Murfad Peer, Mary T. Donofrio and Lasya Gaur
Children’s National Medical Center, Washington, DC, USA
doi:10.1093/icvts/ivt290

© The Author 2013. Published by Oxford University Press on behalf of the European Association for Cardio-Thoracic Surgery. All rights reserved.

We thank Dr Sainathan for his eComment on our article [1, 2]. The author suggests an alternative strategy of placing bilateral branch pulmonary artery bands while leaving the aortopulmonary (AP) window intact as was used by Hosein *et al.* [3] for the management of patients with hypoplastic left heart syndrome (HLHS). While there may be different ways of managing rare complex anatomies such as tricuspid atresia and aortopulmonary window, in addition to the surgical strategy, we once again would like to highlight the interesting physiology in this patient, which we believe is related to ventricular volume unloading following AP window ligation leading to restriction at the level of the ventricular septal defect.

We have successfully used bilateral branch pulmonary artery bands as a bridge to decision in high-risk patients with HLHS [4]. While this approach is to be recommended in certain selected patients, it is not a procedure free of problems, as Hosein *et al.* themselves in their report have accepted as ‘fundamentally flawed’ [2].

Conflict of interest: none declared

References

- [1] Peer SM, Donofrio MT, Gaur L, Sinha P. Tricuspid atresia with aortopulmonary window: challenges in achieving a balanced circulation *Interact CardioVasc Thorac Surg* 2013;17:441–3.
- [2] Sainathan S. eComment. Balancing the pulmonary circulation in tricuspid atresia with an aortopulmonary window. *Interact CardioVasc Thorac Surg* 2013;17:443.
- [3] Hosein RB, Mehta C, Stickley J, Mcguirk SP, Jones TJ, Brawn WJ, Barron DJ. Creation of aorto-pulmonary window with pulmonary artery band is not good palliation for hypoplastic left heart syndrome. *Eur J Cardiothorac Surg* 2007;32:745–50.
- [4] Kurkluoglu M, Jonas RA, Sinha P. Bilateral branch pulmonary artery banding as a bridge to decision/preoperative optimization of high-risk neonates with hypoplastic left heart syndrome. *World J Pediatr Congenit Heart Surg* 2013;227–8.