14. Waterbolk TW, Talsma MD, Loeff BG, Slooff MJ, Ebels T. Increasing cyanosis after total cavopulmonary connection treated by banding a separate liver vein. *Ann Thorac Surg.* 1995;59:1226-8.
15. Burstein DS, Mavroudis C, Puchalski MD, Stewart RD, Blanco CJ, Jacobs ML. Pulmonary arteriovenous malformations in heterotaxy syndrome: the case for early, direct hepatic vein-to-azygos vein connection. *World J Pediatr Congenit Heart Surg.* 2011;2:119-28.
16. Gessler P, Schmitt B, Prêtre R, Latal B. Inflammatory response and neurodevelopmental outcome after open-heart surgery in children. *Pediatr Cardiol.* 2009;30:301-5.
17. Tárnok A, Schneider P. Pediatric cardiac surgery with cardiopulmonary bypass: pathways contributing to transient systemic immune suppression. *Shock.* 2001;16(Suppl 1):24-32.
18. Gaynor JW, Nicolson SC, Jarvik GP, Wernovsky G, Montenegro LM, Burnham NB, et al. Increasing duration of deep hypothermic circulatory arrest is associated with an increased incidence of postoperative electroencephalographic seizures. *J Thorac Cardiovasc Surg.* 2005;130:1278-86.
19. Sakamoto T, Kurosawa H, Shin'oka T, Aoki M, Isomatsu Y. The influence of pH strategy on cerebral and collateral circulation during hypothermic cardiopulmonary bypass in cyanotic patients with heart disease: results of a randomized trial and real-time monitoring. *J Thorac Cardiovasc Surg.* 2004;127:12-9.
20. Morales D, Williams E, John R. Is re sternotomy in cardiac surgery still a problem? *Interact Cardiovasc Thorac Surg.* 2010;11:277-86.
21. Kirshbom PM, Myung RJ, Simsic JM, Kramer ZB, Leong T, Kogon BE, et al. One thousand repeat sternotomies for congenital cardiac surgery: risk factors for reentry injury. *Ann Thorac Surg.* 2009;88:158-61.
22. de Zélicourt DA, Haggerty CM, Sundareswaran KS, Whited BS, Rossignac JR, Kanter KR, et al. Individualized computer-based surgical planning to address pulmonary arteriovenous malformations in patients with a single ventricle with an interrupted inferior vena cava and azygos continuation. *Thorac Cardiovasc Surg.* 2011;141:1170-7.
23. Sundareswaran KS, de Zélicourt D, Sharma S, Kanter KR, Spray TL, Rossignac J, et al. Correction of pulmonary arteriovenous malformation using image-based surgical planning. *JACC Cardiovasc Imaging.* 2009;2:1024-30.
24. Amodeo A, Di Carlo D, Grigioni M, De Santis M, Di Donato RM. Early primary Kawashima operation combined with direct hepatic vein-to-azygos vein connection: a new logical approach. *J Thorac Cardiovasc Surg.* 2005;129:949-50.
25. Sathe Y, Chidambaram S, Manohar K, Cherian KM. Staged Kawashima operation with cavopulmonary connection. *J Thorac Cardiovasc Surg.* 2012;144:267-8.
26. Mondésert B, Marcotte F, Mongeon FP, Dore A, Mercier LA, Ibrahim R, et al. Fontan circulation: success or failure? *Can J Cardiol.* 2013;29:811-20.
27. Anderson PA, Sleeper LA, Mahony L, Colan SD, Atz AM, Breitbart RE, et al. Contemporary outcomes after the Fontan procedure: a Pediatric Heart Network multicenter study. *J Am Coll Cardiol.* 2008;52:85-98.
28. Jahangiri M, Kreutzer J, Zurakowski D, Bacha E, Jonas RA. Evaluation of hemostatic and coagulation factor abnormalities in patients undergoing the Fontan operation. *J Thorac Cardiovasc Surg.* 2000;120:778-82.
29. Uemura H, Ho SY, Devine WA, Kilpatrick LL, Anderson RH. Atrial appendages and venoatrial connections in hearts from patients with visceral heterotaxy. *Ann Thorac Surg.* 1995;60:561-9.

Key Words: Fontan procedure, congenital heart defect, collateral circulation, thoracotomy

EDITORIAL COMMENTARY

When is palliation permanent?

Peter B. Manning, MD

See related article on pages 1524-30.

I recall from early in my training a mentor routinely referring to staged reconstruction for single ventricle cardiac defects as “permanent palliation,” implying that although complete cavopulmonary connection may afford a physiologic correction with separation of the systemic

and pulmonary circulations, there remained long-term liability inherent with the lack of a pulmonary ventricle. In 1984, Kawashima and associates¹ described an experience of 4 patients with “presently uncorrectable cyanotic cardiac anomalies.” Patients with heterotaxia and anomalies of systemic venous drainage were at the time thought to be impossible to manage in the manner that was being proposed for other single ventricle anomalies. They described a “total cavopulmonary shunt operation” for patients with azygos or hemiazygos continuation of the absent inferior vena cava that became eponymously named the “Kawashima” operation. Although they recognized this operation did not constitute a complete diversion of systemic venous return to the pulmonary arteries, leaving the “hepatocardiac venous and coronary sinus flow” to continue to bypass the lungs, they anticipated that this would result in a

From the Division of Cardiothoracic Surgery, St Louis Children's Hospital, St Louis, Mo.

Disclosures: Author has nothing to disclose with regard to commercial support.

Received for publication March 5, 2015; accepted for publication March 6, 2015; available ahead of print April 11, 2015.

Address for reprints: Peter B. Manning, MD, Division of Cardiothoracic Surgery, St Louis Children's Hospital, St Louis, MO 63110 (E-mail: manningp@wudosis.wustl.edu).

J Thorac Cardiovasc Surg 2015;149:1530-1

0022-5223/\$36.00

Copyright © 2015 by The American Association for Thoracic Surgery

<http://dx.doi.org/10.1016/j.jtcvs.2015.03.009>

satisfactory end state, or permanent palliation, for this patient cohort.

The predictable development of pulmonary arteriovenous malformations and the resultant cyanosis and decline in functional status due to the lack of the unidentifiable “hepatic factor” bathing the lungs have led to the recognition that in most patients the Kawashima operation is, in fact, not an equivalent permanent palliation to the more conventional Fontan operation and its modifications. If we agree that the answer to the question, “Is the Kawashima operation a permanent palliation for this group of complex patients with single ventricles?” is “No,” then this raises 2 additional important questions: “What is the best way to enable this hepatic factor to bathe the lungs?”, and “What should be the proper timing of such a maneuver?” The article by Arrigoni and colleagues² helps to address the first question well and shares the authors’ bias regarding the second. There are many disadvantages in simply performing a “routine” completion Fontan operation in this patient population, including the anatomic challenges posed by the often multiple sites of connection of the hepatic veins to the atrium and the difficulty that accompanies routing blood flow from the level of the diaphragm to the pulmonary arteries in the presence of atrial and ventricular situs abnormalities. The lower volume of flow compared with a typical Fontan pathway, in addition to the challenging geometry, likely increases the risk of pathway thrombosis, a complication I have unfortunately seen in my personal experience. The concept of performing an extra-anatomic connection of the hepatic veins to the azygos or hemiazygos vein was first described by Steinberg and colleagues³ in 2003 using a sternotomy approach and cardiopulmonary bypass support. The Groningen team reported applying this concept via thoracotomy, avoiding cardiopulmonary bypass in 2008.⁴ They advocated multiple advantages of this technique, including avoidance of a re sternotomy, avoidance of cardiopulmonary bypass support, and creation of a short prosthetic communication. Their current communication is important because with follow-up to 10 years, they demonstrate the durability of this technique in relieving cyanosis and improving cardiac functional status.² Although the technique they describe is technically simple, their experience

underscores the importance of adequately defining the often unusual hepatic venous connection to the atria when planning for such an intervention via a thoracotomy approach. Despite their practice of routinely obtaining computed tomography angiography to examine this anatomy, 2 of 11 patients required reoperation to manage additional hepatic venous connections that had not been incorporated into their shunt, which thus served as venovenous collaterals leading to persistent cyanosis. As in a game of chess, when one must always be thinking of the next move when planning an attack, examination of the hepatic venous connections at the time of the original Kawashima operation may have supplemented the understanding of this critical anatomy and aided in planning a subsequent hepatic to azygos shunt.

In regard to the proper timing of an intervention to incorporate hepatic venous flow into the cavopulmonary circulation, a number of authors have advocated the routine addition of this step, either as a component of the initial Kawashima operation or in a time frame similar to completion of a traditional Fontan operation, citing the near certainty of the development of pulmonary arteriovenous malformations over time. Arrigoni and colleagues² choose to wait for specific indications of cyanosis or declining functional status. In the absence of any longitudinal follow-up studies of this population examining the actual incidence and liability of pulmonary arteriovenous malformations, their approach is easily defended. Regardless of one’s bias about the timing issue, their experience certainly endorses consideration of this technique for all cases with anatomy amenable to its application.

References

1. Kawashima Y, Kitamura S, Matsuda H, Shimazaki Y, Nakano S, Hirose H. Total cavopulmonary shunt operation in complex cardiac anomalies. A new operation. *J Thorac Cardiovasc Surg.* 1984;87:74-81.
2. Arrigoni SC, van den Heuvel F, Willems TP, Hillege H, Lindberg HL, Berger RMF, et al. Off-pump hepatic to azygos connection via thoracotomy for relief of fistulas after a Kawashima procedure: Ten-year results. *J Thorac Cardiovasc Surg.* 2015;149:1524-30.
3. Steinberg J, Alfieri GM, Brandt B, Smith F, Byrum CJ, Fink GW, et al. New approach to the surgical management of pulmonary arteriovenous malformations after cavopulmonary anastomosis. *Ann Thorac Surg.* 2003;75:1640-2.
4. Lopez FE, van den Heuvel F, Pieper P, Waterbolk TW, Ebels T. Off-pump connection of the hepatic to the azygos vein through a lateral thoracotomy for relief of arteriovenous fistulas after a Kawashima procedure. *Cardiol Young.* 2008;18:311-5.