CONGENITAL HEART DISEASE

Coil occlusion of systemic venous collaterals in hypoplastic left heart syndrome

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Objective: To assess the frequency of systemic venous collaterals to the atria, which may cause desaturation, after stage II reconstructive surgery for hypoplastic left heart syndrome (HLHS) and to determine whether coil occlusion prevents the need for surgical ligation.
Design: Prospective interventional study.
Setting: Tertiary referral centre.
Patients: 27 children with HLHS undergoing cardiac catheterisation between October 1996 and February 2001.
Interventions: 19 children were catheterised prestage II, 1 poststage II, and 17 prestage III. Aortic oxygen saturation (SaAo) and pulmonary artery pressure (pPA) were recorded. Angiography was performed into the left internal jugular vein to look for venous collaterals. If present, they were occluded with Cook MReye coils. Angiography was repeated to confirm occlusion, and SaAo and pPA were remeasured.
Results: Collaterals were found in 7 of 27 children: 1 poststage II and 6 prestage III. These were occluded with 1–3 coils without complication. Mean (SE) SaAo before occlusion was 80.2 (2.1)% in those with collaterals compared with 88.7 (1.0)% in those without (p = 0.007). There was no difference in mean pPA between the two groups. After coil occlusion mean SaAo rose to 83.8 (1.8)% (p = 0.007) and mean pPA rose from 12.5 (1.5) to 14.5 (1.8) mm Hg (p = 0.02). None required surgical ligation.
Conclusion: Angiography should be performed at catheterisation before stage II and III surgery for HLHS to exclude systemic venous collaterals. If present, they may be safely and effectively occluded with coils to improve saturation and prevent the need for subsequent surgical ligation.

The development of reconstructive surgery for hypoplastic left heart syndrome (HLHS) has radically improved the outlook for children with this condition over the past 20 years. HLHS was once universally fatal, but survival figures are now approaching 70% in some centres.[1] [2] Surgery is usually undertaken in three stages: at a few days of age, a few months of age, and 3–4 years, respectively. Stage I, the Norwood procedure, consists of reconstruction of the hypoplastic aorta using the proximal main pulmonary artery, removal of the atrial septum, and insertion of a systemic to pulmonary artery shunt. Stage II, the hemi-Fontan operation, consists of an anastomosis between the superior vena cava (SVC) and the right pulmonary artery, with disconnection of the systemic to pulmonary shunt. Stage III, the Fontan operation, consists of an anastomosis between the inferior vena cava (IVC) and the right pulmonary artery, allowing for complete separation of the pulmonary and systemic circulations.

The hemi-Fontan anastomosis created at stage II is used as primary or intermediate palliation for a wide variety of functionally univentricular hearts.[3] It divides the systemic venous drainage into a higher pressure system (SVC to pulmonary artery) and a lower pressure system (IVC to atria, assuming an unrestrictive interatrial communication). The pressure difference between the two systems is referred to as the transpulmonary pressure gradient. With this pattern of circulation, there may be systemic venous collaterals between the higher pressure SVC–pulmonary artery system and the lower pressure IVC–atrial system, which cause desaturation. These have been shown to occur more frequently in patients with a high transpulmonary pressure gradient in the immediate postoperative period[4] [5] or at follow up catheterisation.[6] However, there is debate as to whether these vessels develop following surgery or whether they are present throughout, opening up in response to the change in pressures.[4–7]

At Guy's and St Thomas' Hospital, cardiac catheterisation is routinely undertaken before stages II and III to assess suitability for surgery. At catheterisation before stage II, one child was found to have an apparently insignificant collateral draining from the innominate vein to the left atrium. However, after surgery her saturations remained low, and a repeat angiogram showed a significant right to left shunt down a greatly enlarged collateral, which required surgical ligation. Following this her saturations improved, and she made an uneventful recovery.

The aim of this study was to assess the frequency of such collaterals in children with HLHS and to determine whether coil occlusion at cardiac catheterisation before surgery prevents the need for subsequent surgical ligation.

PATIENTS AND METHODS

Between October 1996 and February 2001, cardiac catheterisation was performed on 27 children with classic HLHS to assess their suitability for further surgery and to look for collateral vessels. Classic HLHS was defined as a right ventricular dependent circulation in association with atresia or severe hypoplasia of the aortic and mitral valves and the aortic arch, with consequent underdevelopment of the left ventricle. The patency of the left sided valves on the original echocardiogram was noted to see whether there was any correlation with the

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Abbreviations: HLHS, hypoplastic left heart syndrome; IVC, inferior vena cava; pPA, pulmonary artery pressure; SaAo, aortic oxygen saturation; SVC, superior vena cava

Figure 1 Angiogram of hemi-Fontan with venous collateral.

Figure 2 Angiogram of hemi-Fontan with occluded collateral.

presence of collaterals. Nineteen children were assessed before stage II, at a mean (SD) age of 6.6 (1.6) months and mean weight of 6.6 (1.1) kg, and 17 children before stage III, at a mean age of 35.8 (4.5) months and mean weight of 13.2 (1.9) kg. Ten children were catheterised twice during the study period. One further child, who underwent stage II surgery as an emergency without prior catheterisation, was catheterised in the postoperative period because of persisting desaturation.

Each procedure was carried out under a general anaesthetic and baseline conditions of 28% oxygen were maintained. Access was gained through the right or left femoral artery and vein, and left internal jugular vein. As part of each procedure, aortic oxygen saturation (SaAo) and mean pulmonary artery pressure (pPA) were measured and angiography was performed through the left internal jugular vein to look for collaterals. If present, the collaterals were occluded with detachable Cook MReye coils (Cook Inc, Bloomington, Indiana, USA) inserted through the left internal jugular vein. (These are widely available in the UK and are frequently used for arterial duct occlusions. They have the advantage of being easy to control and may be removed if necessary.) Angiography was repeated to confirm occlusion, and SaAo and pPA were remeasured with the coils in situ. Oxygen saturations were measured by pulse oximetry before discharge.

RESULTS

The mean (SE) SaAo before stage II was 77.6 (1.3)% and the mean pPA was 15.9 (1.0) mm Hg. No collaterals were found in the 19 children catheterised before stage II. All of these patients have since successfully undergone the hemi-Fontan operation.

The child who was catheterised postoperatively because of continuing desaturation was found to have a large single collateral arising from the innominate vein, which drained into the coronary sinus. This was successfully occluded with a 5 × 5 and a 3 × 4 coil (coil dimensions are specified first by coil diameter in millimetres and then the number of loops; hence, a 3 × 4 coil has a loop diameter of 3 mm and 4 loops).

Before stage III, the mean SaAo was 86.5 (1.2)% and the mean pPA was 12.2 (0.8) mm Hg. Collaterals were identified in 6 of 17 patients and were successfully occluded with 1–3 coils of varying sizes without complication. Three children were found to have a single vessel arising from the innominate vein draining to the left atrium; in two patients the vessel was occluded with a single coil (5 × 5 and 5 × 4) but the third required two 5 × 5 coils for complete occlusion. In one child, two vessels arose from the cardinal vein, draining to the right atrium, which were occluded with a 3 × 3 and a 3 × 4 coil, respectively. Another child had two collaterals arising separately from the innominate vein, one of which drained to the left atrium and the other to the IVC. These were occluded with a 5 × 4 and a 5 × 5 coil, respectively. The last child had a meshwork of collaterals arising from the innominate vein draining to both atria and the IVC; these were occluded at their origin with three coils: one 5 × 5 and two 3 × 4. Repeat angiography showed complete occlusion in each case. Figures 1 and 2 show angiograms before and after coil occlusion of a large collateral arising from the innominate vein.

Mean SaAo before occlusion was 80.2 (2.1)% in those with collaterals compared with 88.7 (1.0)% in those without (p = 0.007) but there was no significant difference in mean pPA between the two groups. After coil occlusion, mean SaAo rose from 80.2 (2.1)% to 83.8 (1.8)% (p = 0.007), which, though a significant increase, was still considerably lower than the saturations of those without collaterals. This seems to indicate the presence of a small leak through the coils that was not detectable by angiography. However, by the following day there was no difference in peripheral saturation measurement by pulse oximetry between those with collaterals and those without. Mean pPA rose from 12.5 (1.5) mm Hg to 14.5 (1.8) mm Hg following occlusion (p = 0.02).

There did not appear to be any correlation between patency of the left sided heart valves and the presence of collaterals: 4 of 15 children with aortic atresia developed collaterals, as did 3 of 12 with aortic stenosis (p = 0.92), and 4 of 11 children with mitral atresia developed collaterals, as did 3 of 16 with mitral stenosis (p = 0.30).

In total, seven children (all of whom had undergone the hemi-Fontan procedure) were found to have venous collaterals. This gave an overall frequency of 7 of 27 (26%; 95% confidence limits 11% to 46%) or 7 of 18 (39%; 95% confidence limits 17% to 64%) poststage II. Table 1 summarises these results. Of the 10 children who were catheterised twice during the

Table 1 Frequency of collaterals found at cardiac catheterisation before surgical stages II and III in children with hypoplastic left heart syndrome

	Timing of catheterisation			
Patient groups	Prestage II only (n=9)	Poststage II only (n=1)	Prestages II and III (n=10)	Prestage III only (n=7)
Number with collaterals	0	1	1 (prestage III)	5
Number without collaterals	9	0	9	2

study period, none had collaterals prestage II but one was found to have a collateral before stage III. Fourteen children have since successfully undergone the Fontan procedure, including five with occluded collaterals.

DISCUSSION

The data from this study show that systemic venous collaterals draining to the atria (or IVC before stage III) may be a cause of significant desaturation in children undergoing reconstructive surgery for HLHS. This may cause problems in the immediate postoperative period with severe hypoxia. In the longer term it may result in reduced activity levels and increased fatiguability, together with the risks associated with polycythemia. Although this phenomenon has been recognised since cavopulmonary anastomoses were first introduced,[8] relatively little has been written about it, especially within the context of HLHS.

Our results show that the frequency of systemic venous collaterals following hemi-Fontan for HLHS (39%) is slightly higher than that seen following superior cavopulmonary anastomosis for other forms of congenital heart disease: McElhinney and colleagues[4] reported a frequency of 33%, and Magee and associates[5] a frequency of 31%. However, in a recent series of 40 patients, the majority of whom had already undergone the Fontan procedure (with or without prior superior cavopulmonary anastomosis), Weber[6] reported a frequency of 43%. (As might be expected from the circulatory physiology, haemodynamically significant collaterals were more common in patients who had undergone superior cavopulmonary anastomosis, although this difference was not significant.)

As in Weber's series,[6] we found that the majority of collaterals drained to the atria, with only two children having additional vessels draining to the IVC. This differs from previous reports, in which the majority of collaterals drained to the IVC.[4][5] This is relevant because collaterals that drain to the atria create a permanent right to left shunt and should be occluded, whereas those that drain to the IVC are of no significance following the Fontan procedure and may safely be left unless they are large enough to reduce significantly pulmonary blood flow poststage II.

Previous studies have found that the most significant predictive factor for the presence of collaterals was a high transpulmonary pressure gradient either in the immediate postoperative period[4][5] or at follow up catheterisation.[6] At catheterisation before stage III, we found no significant difference in pPA between those with and those without collaterals, suggesting decompression of the SVC system in those with collaterals. Once the collaterals were occluded, the pPA rose significantly.

It is not known whether these venous collaterals are present from birth and become haemodynamically significant only following hemi-Fontan, or whether they develop de novo in response to the changed pattern of circulation and pressures created by surgery. The results from this study seem to support the latter hypothesis, as (a) apart from the index case, no collaterals were found before stage II, and (b) of the 10 children

catheterised twice, one child had a collateral prestage III that was not present prestage II. Three previous studies also found no collaterals before surgery.[4–6] Additionally, the highest reported frequency of collaterals occurred in the oldest patient population, who had had the longest time for them to develop.[6]

However, the two hypotheses need not be mutually exclusive; the fact that profound desaturation may be a problem in the immediate postoperative period suggests that in some cases the existence of these collaterals predates surgery, as there would not be time for them to develop postoperatively.[7] In such cases they may conduct reverse flow in the azygous or hemiazygous systems or result from a reopening of channels present during embryonic development.[5] In view of this, if collaterals are found prestage II, they should be occluded even if they appear to be insignificant.

The seven children who underwent coil occlusion of collaterals in this study have not been found to have any significant further desaturation at follow up clinic visits. However, if the "de novo development" hypothesis is correct, there is a risk that new collaterals may develop once the original collaterals have been occluded. In view of this, any child who becomes significantly more cyanosed at follow up should have repeat angiography to look for new collaterals.

This study shows that angiography into the left internal jugular vein is effective at detecting venous collaterals and may conveniently be performed as part of a diagnostic catheterisation before stages II and III of surgery. These collaterals may be safely and effectively occluded with coils during the procedure to improve saturation and prevent the need for surgical ligation, either during the hemi-Fontan or Fontan procedure or in the postoperative period.

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REFERENCES
1 **Bove EL**. Surgical treatment for hypoplastic left heart syndrome. *Jpn J Thorac Cardiovasc Surg* 1999;**47**:47–56.
2 **Mahle WT**, Spray TL, Wernovsky G, *et al.* Survival after reconstructive surgery for hypoplastic left heart syndrome: a 15-year experience from a single institution. *Circulation* 2000;**102**(19 suppl 3):III136–41.
3 **Karl TR**. Bidirectional cavopulmonary anastomosis [editorial; comment]. *Cardiol Young* 1999;**9**:6–10.
4 **McElhinney DB**, Reddy VM, Hanley FL, *et al.* Systemic venous collateral channels causing desaturation after bi-directional cavopulmonary anastomosis: evaluation and management. *J Am Coll Cardiol* 1997;**30**:817–24.
5 **Magee AG**, McCrindle BW, Mawson J, *et al.* Systemic venous collateral development after the bi-directional cavopulmonary anastomosis: prevalence and predictors. *J Am Coll Cardiol* 1998;**32**:502–8.
6 **Weber HS**. Incidence and predictors for the development of significant supradiaphragmatic decompressing venous collateral channels following creation of Fontan physiology. *Cardiol Young* 2001;**11**:289–94.
7 **Gatzoulis MA**, Shinebourne EA, Redington AN, *et al.* Increasing cyanosis early after cavopulmonary connection caused by abnormal systemic venous channels. *Br Heart J* 1995;**73**:182–6.
8 **Mathur M**, Glenn WWL. Long-term evaluation of cava-pulmonary artery anastomosis. *Surgery* 1973;**74**:899–916.