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Original Article

Bubble contrast echocardiography in detecting pulmonary arteriovenous malformations after modified Fontan operations

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Abstract The development of pulmonary arteriovenous malformations is a well-known complication after Fontan operations, and may result in significant morbidity due to increasing arterial desaturation. We compared the use of bubble contrast echocardiography and pulmonary angiography in detecting such malformations. We also examined which anatomical and haemodynamic variables were associated with their development. Our study includes 20 patients who had undergone modified Fontan procedures, 10 with atriopulmonary and 10 with total cavopulmonary connections, in Gothenburg between 1980 and 1991. All patients underwent cardiac catheterisation and pulmonary angiography. Bubble contrast echocardiography was performed at the same time, with injection of agitated polygelin colloid solution (Haemaccel, Hoechst) into the right and left pulmonary arteries, respectively. Transoesophageal echocardiography was used to detect the appearance of bubble contrast in the pulmonary venous atrium. The aim was also to evaluate the role of hepatic venous blood. Of the 20 patients, 9 (45%) had a positive contrast echocardiography study, compared with only 2 (10%) detected by pulmonary angiography. Patients with positive contrast echocardiography had a significantly lower arterial oxygen saturation than those with negative studies, both at rest (88% vs 95%, $p < 0.01$) and during exercise testing (78% vs 89%, $p = 0.01$). Bubble contrast echocardiography is much more sensitive in detecting pulmonary arteriovenous malformations than pulmonary angiography. By injecting echo contrast into the right and left pulmonary arteries, the method can be made highly selective. Pulmonary arteriovenous malformations develop much more frequently in patients with the Fontan circulation than previously reported.

Keywords: Functionally univentricular heart; cavopulmonary connection; arteriovenous malformations; cross-sectional echocardiography

In 1971, Fontan and Baudet described a technique for successful definite palliation in patients with tricuspid atresia.[1] The cavopulmonary anastomosis, in which the superior caval vein is connected to the pulmonary arteries in the form of a classical or bidirectional Glenn shunt, or the Fontan operation in its various modifications, including the total cavopulmonary anastomosis, remain useful methods of palliation in patients with functionally univentricular hearts. The development of pulmonary arteriovenous malformations is a well-known complication of these procedures, and may result in significant morbidity due to increasing cyanosis.

The prevalence of such malformations in patients with a cavopulmonary anastomosis has not been well documented. Some investigators found them in a quarter of patients with a bidirectional Glenn shunt.[2–4] Their etiology remains unknown, but the abnormal distribution of flow of blood to the lungs, in favour of the lower lobes, after a bidirectional Glenn shunt or a Fontan procedure is considered to be a contributing factor.[2,3] In 1995, Srivastava et al.[4] reported that their development is related to the exclusion of flow from the liver from the pulmonary

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circulation.[4] These findings, in combination with presence of the malformations in patients with hepatic cirrhosis without congenital cardiac disease, have led to the hypothesis that lack of a "hepatic factor" may contribute to their development in patients with hepatic disease, and in some patients after construction of the cavopulmonary anastomosis.[5]

The diagnosis of these malformations has traditionally been made by catheterisation of the right heart combined with and pulmonary angiography.[6] The use of cross-sectional contrast echocardiography for establishing the diagnosis has been described in some case reports.[7–9] Chang et al.[10] described a series of 14 patients with different forms of cavopulmonary anastomosis where they compared bubble contrast echocardiography and pulmonary angiography in detecting the malformations.[10]

The aims of our study were:

- to investigate the prevalence of pulmonary arteriovenous malformations in patients with different forms of cavopulmonary anastomosis,
- to evaluate the role of hepatic venous blood in their development, and
- to compare the sensitivity of contrast echocardiography with that of pulmonary angiography in detecting the malformations.

Materials and methods

Patients

The first Fontan operation in Göteborg was performed in 1980. Between 1980 and 1991, 28 patients underwent Fontan-like procedures, involving construction of atriopulmonary or total cavopulmonary connections, at the The Queen Silvia Children's Hospital, Göteborg University. Of the patients, six died and two patients are living abroad. The remaining 20 patients were included in this study.

There were 10 atriopulmonary and 10 cavopulmonary operations. Initial atriopulmonary connections in two patients had been converted to cavopulmonary anastomosies because of atrial enlargement and arrhythmias. None of the patients has been left with a Glenn shunt as final palliation. Only two patients had their Glenn operation done as a palliation before the Fontan procedure. In both, the superior caval vein had been anastomosed end-to-end with the right pulmonary artery in classical fashion. In all the other 18 patients, the cavopulmonary anastomosis was performed in one stage. A bidirectional Glenn shunt had been constructed in all patients, with the inferior caval vein connected either through a Gore-Tex® tunnel in the right atrium to the pulmonary artery, or by a direct connection from the right atrium to the pulmonary artery. In all patients with isomeric atrial appendages, the venous flow from the liver was directed to the pulmonary circulation through a Gore-Tex® tunnel. Thus, no patient has the hepatic venous blood drained into the systemic circulation.

Clinical characteristics of the 20 patients are summarised in Table 1. The median age at operation was 4.9 years, with a range from 3.0 to 35.0 years. The interval between the cavopulmonary anastomosis and time of study ranged from 7.8 years to 19.8 years, with a median follow-up time of 11.5 years.

We divided the patients into three groups. The first group consists of the two patients in whom a classic Glenn shunt had been constructed. They have the hepatic venous blood drained only to their left lung. The second group contains six patients with isomerism of the atrial appendages with interruption of the inferior caval vein and either azygous or hemiazygous continuation. Of these, five have left and one right isomerism. A bilateral, bidirectional Glenn shunt had been constructed in four of these patients (#4, 5, 6 and 7). Within the overall group, in four the hepatic venous blood drains mainly into the left lung, in one patient it drains solely into the right lung (#6), and in the other patient (#3) symmetrically to both lungs.

The third group represents the remaining 12 patients, who have the hepatic venous blood directed to both lungs.

Although previous studies,[11,12] have documented that, in normal individuals, the bubble contrast does not pass through the pulmonary capillary bed, we recruited an additional 5 patients to serve as controls. These patients had either isolated patency of the arterial duct or an atrial septal defect, defects which were closed with coils or Amplatzer devices in the catheter laboratory.

This study was approved by the Ethics Committee of the Medical Faculty, Göteborg University.

Contrast echocardiography

Transoesophageal echocardiography was performed with Acuson 128XP 10 c echocardiographic equipment. A biplane probe (V510B) was used, and the transducer head was placed in the oesophagus just behind the pulmonary venous atrium. Echocardiographic contrast agent was produced by agitating Haemaccel® manually to form air-filled microbubbles. Haemaccel® has been shown by others to be a safe and reproducible echo contrast agent.[13] One 5 ml syringe and one 2 ml syringe were joined by a three-way stopcock.The 2 ml syringe contained the Haemaccel® and the 5 ml syringe contained 0.2 ml of air. The Haemaccel® was agitated by rapidly flushing the solution back and forth between the two syringes 10 times or more. A mixture of air and

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Table 1. Patient characteristics.

Patient no.	Diagnosis	Type of operation	Previous palliation	Age at surgery (years)	Follow-up time (years)	SaO_2 at rest (%)	SaO_2 at max. load (%)	Hb (g/l)	Isomerism
1	TA	APC	Classic Glenn	15.1	15.1	93	90	127	No
2	TA	APC	Classic Glenn	27.3	19.0	89	78	168	No
3	DIRV, TGA, PA	TCPC	BT	3.0	9.5	96	87	146	Yes
4	DORV, TGA, PS	TCPC	None	7.5	10.4	92	82	161	Yes
5	Unbal. AVSD, SV, TAPVR	TCPC	BT	3.9	11.0	79	63	189	Yes
6	Unbal. AVSD, DORV, TGA	TCPC	BT	5.2	11.8	89	60	209	Yes
7	TGA, PS, LV-hypoplasia	TCPC	BPT, BT	4.0	8.9	87	82	150	Yes
8	TA, PAPVR	APC	BT	13.3	18.8	91	86	151	Yes
9	TA	TCPC**	BT	5.4	9.3	96	85	133	No
10	PAIVS	APC*	BT	4.2	14.2	99	86	135	No
11	PAIVS	TCPC	BT	4.4	7.8	94	86	139	No
12	TA	TCPC	BT	4.5	8.3	98	94	137	No
13	DILV, TGA, VSD	APC	BPT, BT, DKS	4.5	9.5	97	94	149	No
14	TA	APC*	BT	3.1	15.2	89	84	163	No
15	Ebstein's malformation	TCPC	BT	4.6	9.9	95	92	160	No
16	TA,PS	TCPC	None	4.3	8.3	90	85	144	No
17	TA, DOLV, PS	APC	None	35.0	19.2	93	96	161	No
18	Funct. single ven., TGA, PS	APC	BT	25.8	19.3	88	78	181	No
19	Straddl tricusp.v, DORV	APC	BPT	9.8	12.1	89	79	185	No
20	TA	APC	None	20.4	19.8	97	94	158	No

*Reoperation to TCPC; **Reoperation from TCPC constructed from atrial wall to TCPC with a Gore-Tex® tunnel
Abbreviations: APC: atriopulmonary connection; AVSD: atrio-ventricular septal defect; BT: blalock taussig shunt; DILV: double inlet left ventricle; DIRV: double inlet right ventricle; DOLV: double outlet left ventricle; DORV: double outlet right ventricle; DKS: Damus-Kaye-Stansel anastomosis; PA: pulmonary atresia; BPT: banding of pulmonary trunk; PAIVS: pulmonary atresia and intact ventricular septum; Funct.: Functionally; Unbal.: unbalanced; PAPVR: partial anomalous pulmonary venous return; PS: pulmonary stenosis; TA: tricuspid atresia; TAPVR: total anomalous pulmonary venous return; TCPC: total cavopulmonary connection, TGA: discordant ventriculo-arterial connections

liquid was thereby generated. An end-hole or side-hole catheter was placed with its tip in the right and left branches of the pulmonary trunk before their first bifurcation, without wedging in small branches. The site was controlled before each injection of Haemaccel® by injecting angiographic contrast to make sure that there was no backward flow. To maintain an exact speed of the injection, we used angiographic equipment (Medrad, maRK V Plus). We injected 1.6 ml of the agitated Haemaccel® in the catheter, and the angiographic injector delivered the bolus at a speed of 8 ml/sec. Simultaneous trans-esophageal echocardiography was performed to visualise the presence of bubble contrast in the pulmonary venous atrium (Fig. 1). The amount of microbubbles was subjectively graded from mild (+) to moderate (++) or severe (+++). Each injection was recorded on videotape. In a normal individual, after injection of bubble contrast in the pulmonary arteries, the bubbles are lost in the passage through the pulmonary capillary bed, and therefore almost no bubbles could be detected in the "left heart".[12] The presence of bubble contrast in the pulmonary venous atrium and systemic ventricle is, therefore, proof of intrapulmonary arteriovenous shunting.

Pulmonary angiography

All the 20 patients underwent right and left heart catheterisation including pulmonary angiography. The pulmonary arteries were entered in an antegrade manner through the femoral veins. In the two patients with a classic Glenn anastomosis, the right internal jugular vein was used for access to the right pulmonary artery. In the patients with an interrupted inferior caval vein, we reached the pulmonary arteries via the azygous or hemiazygous continuation. The pulmonary arterial pressures, and superior caval venous saturations, were recorded before the angiography. Selective pulmonary angiograms were performed with an angiography catheter and the tip of the catheter was positioned in the right and left

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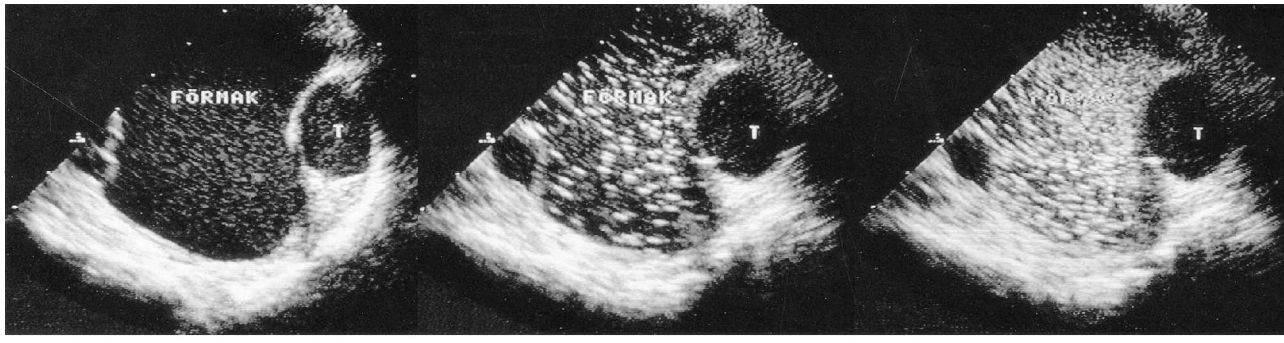

Figure 1.
Contrast echocardiographic study in our fifth patient. (a) Injection of Haemaccel® in the left *pulmonary artery. No microbubbles were detected in the pulmonary venous atrium. (b) The same patient 5 seconds after injection of Haemaccel® in the* right *pulmonary artery. (c) The same recording approximately 5 seconds later. There is complete opacification of the pulmonary venous atrium. (FÖRMAK = atrium, T = tunnel)*

pulmonary arteries, to ensure adequate filling of the contrast, and to avoid wedging in the small branches. The fluoroscopic cameras were angled at straight anterior–posterior and lateral positions to ensure adequate demonstration of the entire hemithorax, including the lower lobes of the lungs. Cineangiograms were recorded for later review. Angiograms were examined blindly by one paediatric cardiologist and one paediatric radiologist for pathological findings indicating the presence of pulmonary arteriovenous malformations.

The distribution of hepatic venous blood flow was examined by injection of angiographic contrast in the lower part of the tunnel or in the right atrium. An example is shown in Figure 2.

Lung perfusion scintigraphy

In order to examine the pulmonary distribution of the hepatic venous return further in the patients with flow of hepatic venous blood to both lungs, we performed lung perfusion scintigraphy with injection of ^{99m}Tc-macroaggregated albumin in a leg in 7 of the 12 patients.

Statistics

Groups were compared using the Mann-Whitney test.

Results

All five patients serving as controls had negative findings, both for contrast echocardiograms and angiographic findings. The results of angiography and contrast echocardiography for those undergoing the Fontan procedures are shown in Table 2. Of the 20 patients, 9 with a cavopulmonary anastomosis had positive contrast echocardiography (45%). In only two cases was there angiographic evidence of pulmonary arteriovenous malformations (10%). Both these patients also had positive contrast echocardiography. On angiography, large vessels were visualised extending distally into the lower zones of the lungs, along with absence of the capillary phase and early filling of the pulmonary veins. Both patients had marked cyanosis at rest, which progressed during exercise, with aortic saturarions of 60%, and 63%, respectively at maximal load.

There was no significant difference in the time of follow-up between the patients with positive contrast echocardiography and those presenting negative studies. Both patients with a classic Glenn shunt had a positive echo contrast study in the lung which did not receive flow from hepatic veins (patients #1 and 2). Of the patients with isomerism, 5 (83%) were positive on contrast echocardiography, and two of these patients were the one with positive angiographic findings. In all of them, the pulmonary arteriovenous malformations were present in the lung with no or minimal hepatic venous flow. Of the 12 patients with hepatic venous blood to both lungs, 2 (17%) were positive on contrast echocardiography, both in their right lung. Lung perfusion scintigraphy showed that, in both these cases, most of the venous return from the lower body was directed to the left lung (75% and 86% respectively).

No patient had pulmonary arteriovenous malformations in both lungs. Patients with pulmonary arteriovenous malformations had significantly lower oxygen saturation compared with patients with negative studies. This was found at rest, 88% vs 95%, $p < 0.01$, and also during maximal exercise, 78% vs 89%, $p < 0.01$. The patients with pulmonary arteriovenous malformations also tended to have higher haemoglobin counts than the patients without

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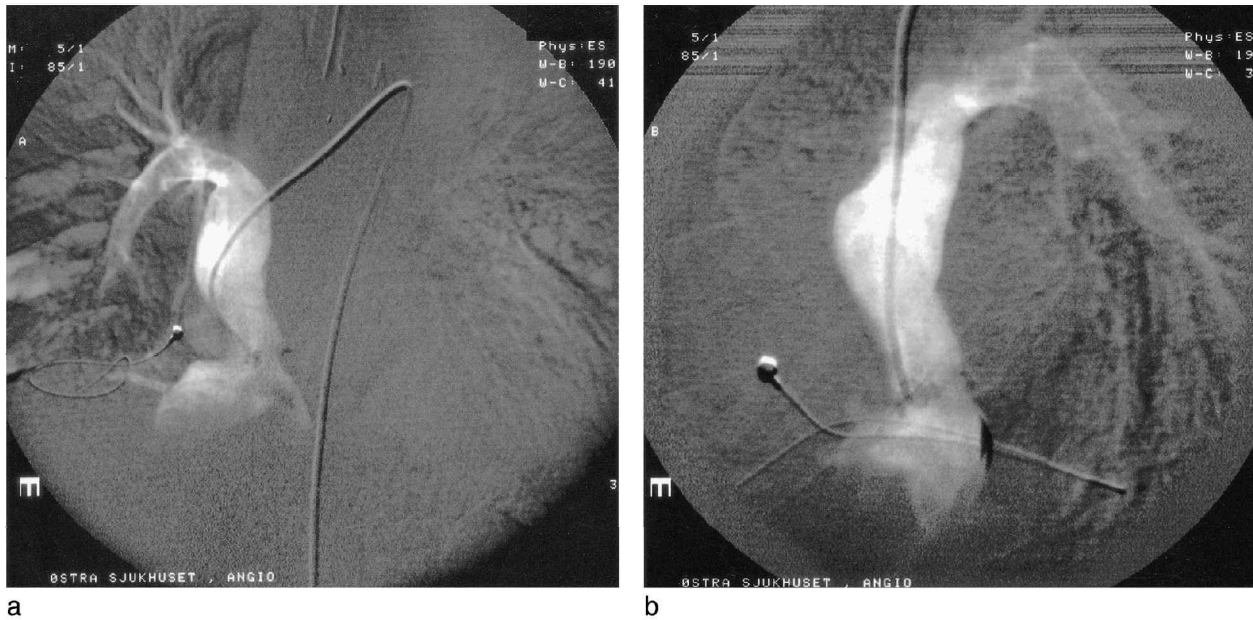

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Figure 2.
Angiography in our sixth patient showing that all flow from the tunnel, that is hepatic venous flow, is distributed solely to the right pulmonary artery. (a) Frontal view; (b) Lateral view.

Table 2. Results of cardiac catheterisation and presence of Pulmonary Arteriovenous Malformations (PAVM) according to angiography and bubble contrast echocardiography as well as results of lung perfusion scintigraphy after isotope injection in a lower extremity.

Patient no.	Mean PAP (mmHg)	SCV sat (%)	SaO_2 (%)	Presence of PAVM bubble contrast echocardiography		Presence of PAVM angiography		Hepatic venous blood flow to pulmonary circulation		Scintigraphic pulmonary blood flow distribution (%)	
				Left	Right	Left	Right	Left	Right	Left	Right
1	11	76	93		+			Yes	No		
2	12	74	89		++			Yes	No		
3	9	80	96					Yes	Yes		
4	7	66	92		+			Yes	No		
5	11	62	79		+++		Pos	Yes	No		
6	13	82	89	+++		Pos		No	Yes		
7	14	79	87		++			Yes	No		
8	14	69	91		++			Yes	No		
9	11	73	96					Yes	Yes	Not done	
10	10	79	99					Yes	Yes	Not done	
11	13	66	94	*		*		*	Yes	Not done	
12	11	75	98					Yes	Yes	38	62
13	14	80	97					Yes	Yes	Not done	
14	8	69	89					Yes	Yes	54	46
15	10	75	95					Yes	Yes	Not done	
16	10	74	90		++			Yes	Yes	75	25
17	13	76	93					Yes	Yes	37	63
18	12	77	88					Yes	Yes	24	76
19	12	62	89		+			Yes	Yes	86	14
20	16	76	97					Yes	Yes	35	65

*Absence of blood flow to the left pulmonary artery (unknown before catheterisation)
(+) mild, (++) moderate, (+++) severe intrapulmonary shunting; SCV: superior caval vein

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intrapulmonary right-to-left shunts, but this difference was not statistically significant.

Discussion

Although the exact prevalence of pulmonary arteriovenous malformations is unknown in patients with the Fontan circulation, Kopf et al.[11] reported their incidence to increase with time after construction of cavopulmonary anastomoses. They found a prevalence of 31% after a mean follow up time of 6.8 years in patients with Glenn shunts. Laks et al.[2] described such malformations in up to one quarter of their patients with Glenn shunts. All these findings, however, were based on angiographic studies.

Chang et al.[10] compared transthoracic contrast echocardiography, using hand-agitated saline, and selective pulmonary angiography in detecting the malformations, finding 71% to be positive with contrast echocardiography, as opposed to only 21% using angiography. We also compared contrast echocardiography with selective pulmonary angiography, and our findings are in keeping with those of Chang and colleagues.[10]

In a normal individual, the microbubbles will be lost in the pulmonary circulation and animal studies have shown that the lungs are superb filters for air bubbles. The bubbles will only escape entrapment by the pulmonary capillary system if the lungs are overloaded with more than 20 ml of air.[12,14] We used only 0.2 ml of air in each injection of bubble contrast. It is unlikely, therefore, that our findings are false positive. Furthermore, no patient in our study serving as a control had any return of microbubbles from the pulmonary circulation.

We demonstrated, therefore, that contrast echocardiography is much more sensitive in detecting pulmonary arteriovenous malformations than pulmonary angiography.

The two patients in our study who were positive on angiography had a very large amount of microbubbles in the pulmonary venous atrium on contrast echocardiography. They were also deeply cyanotic and had a high haemoglobin count, indicating massive pulmonary arteriovenous malformations.The patients who were positive on contrast echocardiography but negative on angiography were probably in an earlier stage of development of the malformations. We found that those with the malformations had a significantly lower arterial oxygen saturation at rest, and a more pronounced decrease in saturation during exercise, compared to those without malformations. Hence, contrast echocardiography can be used for screening patients so as to detect early stages of formation of the lesions, especially in patients with arterial desaturation at rest and/or during exercise.

By injecting Haemaccel® into branches of pulmonary arteries, the method can also be made very selective. This is especially important if the choice of treatment is embolisation.

We found that, in patients with no or minimal hepatic venous return to one lung, all but one lung showed positive contrast echocardiographic findings. In the two patients with hepatic venous blood to both lungs and positive echo contrast studies, scintigraphy showed that almost all flow from the lower body was directed to the lung without pulmonary arteriovenous malformations. Thus, although the so-called "hepatic factor" remains to be indentified, our results provide strong evidence for lack of hepatic venous blood flow to the lungs as a major factor contributing to the development of pulmonary arteriovenous malformations in patients with the Fontan circulation.

There are some case reports suggesting that the malformations can be reversed. Graham et al.[15] reported on a patient with a functionally univentricular heart and left isomerism in whom, after construction of a bilateral bidirectional Glenn shunt, all the hepatic venous blood drained directly into the systemic circulation. The patient developed progressive cyanosis due to pulmonary arteriovenous malformations, together with a deterioration in ventricular function. Fourteen months after cardiac transplantation, resolution of the malformations was complete. Similarly, Shah et al.[16] described 3 patients with isomerism who developed pulmonary arteriovenous malformations after construction of a superior cavopulmonary connection, with resolution after inclusion of hepatic veins in the pulmonary circulation. In a recent paper, a surgical approach resulting in diversion of the hepatic venous blood flow to the lungs in a balanced fashion has been advocated to avoid the development of pulmonary arteriovenous malformations.[17]

In conclusion, therefore, pulmonary arteriovenous malformations can be detected by contrast echocardiography after construction of the Fontan circulation in many more patients than suggested by previous reports based on angiography. Lack of hepatic venous blood flow to the pulmonary circulation is probably the most important factor for the development of these malformations. Children with the Fontan circulation should be screened for pulmonary arteriovenous malformations with contrast echocardiography if they develop falling saturation at rest, or if there is a significant drop in saturation during exercise testing. No patient should be left with the hepatic venous blood draining to the systemic circulation, or with an assymmetric distribution of the hepatic venous blood flow between the lungs.

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