Results after Norwood procedure and subsequent cavopulmonary anastomoses for typical hypoplastic left heart syndrome and similar complex cardiovascular malformations

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Abstract

Objective: From October 1989 to June 1998, 60 patients have undergone the Norwood procedure (stage I) at our institution. The results of the staged surgical reconstruction and risk factors were analyzed. Typical hypoplastic left heart syndrome (HLHS) and complex lesions with aortic hypoplasia were compared with each other.

Patients: Typical HLHS: N = 48, median age 15 days (5–190 days), median weight 3.6 kg (2.6–5.3 kg). Complex lesions (dominant left ventricle): N = 12, median age 59 days (10–884 days), median weight 3.4 kg (2.4–12 kg).

Results: Typical HLHS: The stage-I hospital survival was 73% (35/48). It improved from 60% (95% confidence interval: 49–71%) during 1989–1994 to 91% (95% CI: 81–100%) during 1997–1998. Seven patients were lost late. The right ventricular end diastolic diameter (P = 0.015), shortening fraction (P = 0.027), and the presence of an obstructed pulmonary venous return (P = 0.0032) were significant risk factors. 23 children underwent stage-II operation with four (17%) deaths. All survivors experienced an improvement of their statomotoric development. Stage-III operation was performed in 13 patients with no hospital death. Follow up after stage-III procedure was 7 months to 7 years. 4 year actuarial survival, including hospital mortality and deaths at subsequent stages, improved from 28% (95% CI: 18–38%) during 1989–1994 to 58% (95% CI: 48–68%) during 1994–1997. No patient had signs of myocardial insufficiency. Complex lesions: Stage-I hospital survival was 83% (10/12) with no late death. Stage-II was performed in 8 and stage-III in 6 patients with no death respectively.

Conclusion: In typical HLHS the results have improved over time. Both size and function of the right ventricle determined significantly stage-I survival. An early operation prevents the natural progression of pulmonary blood flow and may weaken all three risk factors. Patients with complex lesions seemed to have better chances of surviving the early postoperative period. The multistage reconstructions have become a realistic option for patients with HLHS and similar lesions, regardless of the morphologic subtype or diminutiveness of the aorta, and lead to an acceptable quality of life.

Keywords: Hypoplastic left heart syndrome; Norwood procedure; Risk factors; Single ventricle; Fontan operation; Total cavopulmonary anastomosis

1. Introduction

Typical hypoplastic left heart syndrome (HLHS) is the most common defect with one functional ventricle. Without any therapy 95% of all infants with HLHS do not survive the first month of life. Since October 1989 we have offered the staged reconstructive procedure to palliate neonates with typical HLHS as well as patients with complex cardiovascular malformations associated with hypoplastic aortic orifice and aortic arch. The main difference between both types of cardiovascular lesions is the systemic ventricle. It is a morphological right chamber in typical HLHS and a morphological left chamber in the cases with complex lesions. The results of the Norwood and subsequent Fontan procedures for typical HLHS were reviewed over three consecutive time periods and potential risk factors for early or late cardiac death after the Norwood procedure were analysed. The results of the Norwood and subsequent Fontan procedures for complex cardiovascular malforma-
tions were reviewed separately and were compared with typical HLHS. Since the last group consisted of 12 patients only we did not subdivide the whole period into several consecutive time periods, nor did we analyse risk factors.

2. Patients and methods

2.1. Patient data

2.1.1. Typical HLHS

From October 1989 to June 1998, 48 infants underwent stage-I reconstruction (Norwood procedure) eventually leading to repair by total cavopulmonary anastomosis. The median age was 15 days (range 4–190 days), the median weight was 3.6 kg (range 2.6–5.4 kg). From 1989 to 1994 all newborns with the diagnosis of typical HLHS were offered the stage-I reconstruction (Norwood procedure) and none was excluded. The smallest neonate we have operated upon was 2.6 kg. Between 1994 and 1997 we excluded two neonates with a weight of less than 2.6 kg since at that time we were worried about the length of the circulatory arrest and about sufficient cerebral protection. However, in the meantime our experience has increased and the weight limit is disaffirmed since 1997. Since 1994 two further neonates were not accepted for operation. Both patients had experienced a pH of 6.9 for at least 1 h in the referring hospital and both were cardiopulmonary resuscitated. Normal brain and myocardial function was dubious.

Each patient had a complete examination with Doppler echocardiography for non-invasive evaluation of cardiac anatomy. The echocardiographic diagnosis of typical HLHS was established if all three segments of the left ventricle were underdeveloped. Cases with patent aortic or mitral valves were taken into the series if at least two of the following echocardiographic criteria were met: (1) aortic annulus diameter of 5 mm or less, (2) mitral annulus diameter of 7 mm or less, and (3) a LV/RV length ratio of 0.8 or less. The one infant with atrioventricular septal defect had a small left ventricle and only a quarter of the common atrioventricular valve was committed to the left ventricle. In addition there was coarctation of the aorta. Before stage-I procedure angiocardiography was performed only in the few cases with a borderline sized left ventricle. However, angiocardiography was routinely performed before stage-II and stage-III procedures.

To estimate the degree of ventricular dilatation the upper double standard deviation of the normal mean value of the right or the left ventricular end diastolic diameter corresponding to each patient’s body size [1] was defined as the patient’s upper normal limit. This was set as 100%. The ventricular diameters were expressed in per cent of the upper normal limit. The right ventricular shortening fraction was measured in a modified short axis view below the opened leaflets of the tricuspid valve.

![Table 1](image)

Table 1
Diagnoses of the 12 complex cardiovascular malformations with hypoplasia or atresia of the aorta (aa ascending aorta, a.arch aortic arch, DILV double inlet left ventricle, DORV double outlet right ventricle, IAA-B interrupted aortic arch type B, RV right ventricle, TGA transposition of the great arteries, VSD ventricular septal defect)

<table>
<thead>
<tr>
<th>n</th>
<th>Diagnosis</th>
<th>Annulus diameter</th>
<th>Length ratio</th>
</tr>
</thead>
<tbody>
<tr>
<td>3</td>
<td>DILV, I-TGA, RV-hypoplasia, VSD restrictive, aa 2–4 mm</td>
<td>5 mm</td>
<td>0.8 or less</td>
</tr>
<tr>
<td>3</td>
<td>TCA, I-TGA, RV-hypoplasia, VSD restrictive, aa 2–4 mm, IAA-B</td>
<td>3 mm +</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>DILV, I-TGA, RV-hypoplasia, VSD restrictive, aa 3 mm + IAA-B (n = 1) and aa 5 mm + a.arch 4 mm (n = 1)</td>
<td>3 mm +</td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>DORV, RV-hypoplasia, subaortic stenosis, a.arch 4 mm</td>
<td>3 mm</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>two ventricles, VSD, aortic atresia, aa &lt;2 mm, IAA-B</td>
<td>3 mm</td>
<td></td>
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Fig. 1. Angiography of a newborn with two equal ventricles, VSD, atresia of the aortic orifice, hypoplastic ascending aorta and interruption of the aortic arch type B. The thoracic aorta is supported by a persistent left ductus arteriosus and the ascending aorta and the coronary arteries are supported by a persistent right ductus arteriosus. The catheter is placed within the right and the left ductus, respectively.
2.1.2. Morphologic characteristics of the typical HLHS

Aortic and mitral atresia: 11 patients (23%). Two had in addition unilateral renal agenesis. Aortic atresia and mitral hypoplasia: 8 patients (16%). Aortic hypoplasia and mitral atresia: 9 patients (19%). One of them had an associated stenotic supracardiac total anomalous pulmonary venous connection. Aortic and mitral hypoplasia: 19 patients (40%). One had in addition hypoplastic right lung syndrome. Hypoplastic left ventricle with complete atrioventricular septal defect: 1 patient (2%).

The mean diameter of the ascending aorta was $3.7 \pm 2.3$ mm (range 1.5–10 mm). It was <2 mm in 11 patients (23%), 2 to 3 mm in 13 patients (27%), 3–4 mm in 7 patients (16%), and >4 mm in 17 patients (35%).

2.1.3. Complex lesions

During the same 8.75 years, 12 patients with complex cardiovascular malformations underwent stage-I reconstruction (Norwood procedure). The detailed diagnoses are summarized in Table 1. Nine patients had a hypoplastic subaortic right ventricle and a hypoplastic aorta. Three patients had two ventricles with a VSD, an aortic atresia, a diminutive ascending aorta, an interrupted aortic arch type B with one or bilateral ductus arteriosi (Fig. 1). All 12 patients had a left ventricle. The diameter of the aortic arch was below 5 mm or absent in 9 cases. A Prostaglandin $E_1$ infusion for sufficient systemic perfusion was necessary in 7 patients. Median age at operation was 59 days (range 10 to 884 days), median weight was 3.4 kg (range 2.4–12 kg).

2.2. Methods

Concerning survival after stage-I procedure of typical HLHS the whole period was divided into three time periods. During the first time period (1989 to 1994) the operative technique varied several times. Since July 1994 the technique was rather standardized. In October 1997 the management during the rewarming period concerning bleeding management, ventilation, volume supply, and balancing pulmonary and systemic resistances was reviewed and partially changed. Since these measures significantly influenced the results, a third time period from November 1997 until June 1998 was established.

2.2.1. Surgical technique

During the first time period the techniques of reconstructing the aortic arch and dissecting of the pulmonary arteries, as well as type, size, and location of the aortopulmonary anastomosis were varied several times. But since 1994 the method of surgical repair has remained essentially constant and is very similar to the technique described in detail elsewhere [2–4]. Following median sternotomy and pericardiotomy the persistent ductus arteriosus is directly cannulated without clamping of the fragile ductus wall tissue through an appropriate small incision in a pouch like suture. After institution of the extracorporal circulation the ductus is ligated distal to the pulmonary artery bifurcation. A single atrial cannula drains the venous blood. Mean cooling time is 25 min, neither shorter than 20 min nor longer than 30 min. No cardioplegia is used. At the beginning of hypothermic circulatory arrest the body temperature is 15–17°C. The heart lung machine is primed to produce a haematocrit of 20%. The acid base status is controlled by pH-stat system. After complete atrioseptectomy has been performed the pulmonary artery is transected above the pulmonary valve commissures. Distal to the pulmonary artery bifurcation the ductus arteriosus is ligated with a silk suture. The ductus is transected distal to the silk suture and followed to the descending aorta. The descending aorta is dissected free 1 cm distal to the ductus entry and the ductus excised from its entry into the descending aorta. At its concavity the aortic arch is then incised longitudinally. The length of incision extends to the level of the transected pulmonary artery. We use a tailored segment of a 10 mm coated Heamashield dacron double-velours prosthesis to augment the hypoplastic aortic arch. The prosthesis is sutured to the aortic arch beginning at the distal end of the incision up to the level of the transected pulmonary artery, using a continuous 6.0 prolene suture. The ascending aorta is connected laterally with interrupted 7.0 prolene sutures to the transected central pulmonary artery before the prosthesis is anastomosed to the main pulmonary artery (neoaorta). Since 1994 a modified Blalock Taussig shunt is used routinely from the innominate artery or from the neoaortic arch to the pulmonary artery close to the bifurcation. After restarting the extracorporal circulation the perfusate temperature is kept at 12°C for the first 10 min. Then gradual rewarming follows with a blood–body gradient of 8°C. Sodium nitroprussid and phosphodiesterase inhibitor is given for high systemic vascular resistance. The Blalock Taussig shunt is kept occluded with a soft clamp till the body temperature reaches 34°C and ventricular ejection is sufficient. Thereafter the Blalock Taussig shunt is opened and ventilation is restarted. When coming off bypass the balance between systemic and pulmonary perfusion is regulated according to arterial blood pressure, pH, $\text{paO}_2$, $\text{paCO}_2$, systemic venous $\text{SO}_2$. This is achieved by varying FiO$_2$, volume supply, application of additional CO$_2$, antagonizing metabolic acidosis, and inotropic support if necessary. A $\text{SaO}_2$ between 70 to 80% and an aortic diastolic pressure of at least 30 mmHg are aspired. The thorax is temporarily closed with a heterologous pericardial patch. Definitive closure of the chest is performed some days later.

For the upper bidirectional cavopulmonary anastomosis the neoaorta and the modified Blalock-Taussig shunt are exposed and the neoaorta is cannulated. The superior vena cava is cannulated at the level of the innominate vein and with normothermic extracorporal perfusion it is transected at the level of the right pulmonary artery. The proximal stump is closed with continuous 6.0 prolene suture. The right pulmonary artery is incised longitudinally according to the width of the superior vena cava. Following an oval
excision of the pulmonary artery wall an end to side anastomosis is accomplished with a 6.0 prolene suture. The ventricle is gradually loaded and finally extracorporeal circulation is stopped.

For the total cavopulmonary anastomosis the cannulation of the aorta and the superior vena cava are performed as described above. The inferior vena cava is cannulated through the right atrium as far laterally and caudally as possible. With total extracorporeal circulation, mild hypothermia, unclamped aorta and ventricular fibrillation, the right atrium is opened. The incision extends cranially and laterally to the closed superior caval stump. The vena cava stump is then anastomosed end to side to the inferior edge of the central portion of the right pulmonary artery. The anastomosis is augmented by a tailored segment of coated Heamashield Dacron Double velours prosthesis. The intra-atrial tunnel is always fenestrated with a 4 mm punch. When the patient is normothermic the right atrium is closed and extracorporeal circulation is stopped. A catheter is placed into the left atrium. Bilateral pleural drainage are inserted if possible.

2.2.2. Surgical data

The mean circulatory arrest time was 68 ± 15 min. In the last 36 patients a modified Blalock-Taussig shunt was used with 4 mm in 29 patients, a 3.5 mm shunt in 6 patients, and a 3 mm shunt in one patient. The length of the shunts varied between 18 and 35 mm. Mean circulatory arrest time of patients with complex cardiovascular lesions was 36 ± 17 min. A 4 mm Blalock Taussig shunt was used in 10 cases and a 3.5 mm shunt in 2 patients respectively.

2.2.3. Data collection and statistics

The medical records, operative reports and echocardiographic results for all patients were reviewed. Subsequent operative procedures were noted. Hospital mortality was defined as death within the first 30 postoperative days. Follow-up is 100% complete.

For risk factor analysis, all deaths were taken into account except the three deaths due to infections and the two transplanted patients. The following items were evaluated as potential risk factors for death: age, weight, initial pH at the time of admission, average arterial oxygen saturation of the 2 days before operation, preoperative tricuspid incompetence, morphologic subgroup, diameter of ascending aorta, cardiothoracic ratio, right ventricular end diastolic diameter (in relation to normal values corrected for body size), shortening fraction of the right ventricle, presence of an obstructed pulmonary venous return, and intraoperative circulatory arrest time. Survival analysis was performed with the Kaplan–Meier method, univariate risk factors were determined with log rank test. Cox, regression models were defined for multivariate analysis of survival risk factors. In the multivariate analysis, all univariate risk factors appeared to be independently significant. Significance was stated when P-values were <0.05. For all analyses a commercially available standard software package (SPSS Version 8.0, SPSS Inc., Chicago) was used.

3. Results

3.1. Echocardiographic data:

3.1.1. Typical HLHS

The end diastolic diameter of all the 48 right ventricles measured before the Norwood procedure was bigger than normal. The mean value was 180 ± 48% of the upper normal limit (Fig. 2). Stage-I survivors (n = 28) had a right ventricular end diastolic diameter of 166 ± 38% of the upper normal limit and stage-I non-survivors of 208 ± 52% (P < 0.05) respectively (n = 20; included are the 4 deaths after stage-II and excluded are the 3 deaths due to infections and the 2 transplanted patients). The mean right ventricular shortening fraction was 28 ± 8%. Survivors had a shortening fraction of 32 ± 6% and non-survivors of 24 ± 5% respectively (P < 0.05). An obstructed pulmonary venous return was present in 14 patients (13 restrictive foramen ovale and one stenotic total anomalous pulmonary venous connection). The right ventricular end diastolic diameter was neither significantly correlated with the preoperative arterial oxygen saturation nor with the presence of a preoperative tricuspid incompetence.

3.1.2. Complex lesions

The left ventricle of the 12 patients had an end diastolic diameter of 108 ± 31% of the upper normal limit before the Norwood procedure (Fig. 2) and left ventricular shortening fraction was 33 ± 8%.

3.1.3. Survival: typical HLHS

The reported period of 8.75 years includes the complete learning curve. Table 2 summarizes the results of all operations including cardiac and non-cardiac deaths and two transplantations after stage-I.
Hospital mortality after stage-I procedure was 27% (n = 13, 95% confidence interval: 16–38%). The three main reasons for an early postoperative death were aortopulmonary shunts that were too small, particularly during the first time period, myocardial failure, and intractable bleeding. Two patients died after secondary closure of the chest despite good cardiopulmonary function at the 11th and 12th day respectively. 7 patients were lost late: Two had cardiomegaly in addition to arch obstruction and died due to myocardial failure, one died during surgical revision of a thrombotic BT-shunt 7 weeks after stage-I procedure, 2 patients died due to recurrent infections, both infants suffered from severe DiGeorge syndrome. The tricuspid valve improved from 28% (95% CI: 18–38%) during the first time period to about 10% (1 out of 11 patients, 95% CI: 0–20%) during the last time period. The 4 year actuarial survival improved from 28% (95% CI: 18–38%) during the first time period to about 58% (95% CI: 48–68%) during the second time period.

3.1.5. Risk factor analysis

Risk factor stratification for typical HLHS identified three independent risk factors for stage-I mortality in multivariate analyses: (1) right ventricular end diastolic diameter corrected for body size in percent of the upper normal limit: patients with a right ventricular end diastolic diameter of less than 180% of the upper normal limit had a better chance of survival (P = 0.015) than patients with bigger hearts; (2) right ventricular shortening fraction: patients with a right ventricular shortening fraction of 28% or more had a better chance of survival (P = 0.027) than those with a shortening fraction of less than 28%; (3) the presence of an obstructed pulmonary venous return (P = 0.0032). All the other parameters were not found to be predictors of mortality.

No patient had the combination of all three risk factors, 16 patients had two risk factors, 21 patients had one risk factor, and 11 patients had none of the above mentioned risk factors. Fig. 5 shows the probabilities of early and late survival after stage-I operation for patients with none, with one, and with two risk factors in each of the three time periods. In all three time periods patients with two risk factors had the worst outcome. However, probability of survival improved with increasing experience even for that subgroup.

3.1.6. Follow up after stage-I

Six patients had developed tricuspid incompetence of moderate to severe degree. Four of them had a severely dilated right ventricle and died after stage-II operation. Their end diastolic diameter before stage-II was 230 ± 40% of the upper normal limit. The angiocardiology of those patients performed before stage-II operation had documented an unobstructed aortic arch. The tricuspid incompetence of the remaining 2 patients improved to mild degree after stage-II operation.

Reobstruction of the aortic arch occurred in 10 patients: Four of them were already mentioned above (2 late deaths, 2 transplanted patients). In the remaining cases the obstruction was relieved surgically in 5 and by balloon dilatation in one. The last 6 patients survived and underwent stage-II and eventually stage-III successfully.

After stage-I reconstruction mean arterial oxygen saturation was 82% (range 76%–91%). Four patients underwent early stage-II reconstruction due to hyperperfusion of the lungs at the age of 8 to 12 weeks.
3.1.7. Fontan operation

23 out of the 28 stage-I long-term survivors underwent the bidirectional cavopulmonary anastomosis at a median age of 5 months (2–14 months). The remaining 5 patients are still awaiting the second step. 19 patients (83%) survived the operation. As already mentioned above there were 4 non-survivors who all had suffered from severe right ventricular dilatation and severe tricuspid incompetence prior to the operation. Before stage-III procedure mean arterial oxygen saturation of the 19 survivors was 81 ± 2%. Until stage-III these infants experienced a significant improvement of their statomotoric development and their weight gain. They learned to sit and to stand and the body weight, which was below the 3rd percentile before stage-II procedure in each case, was found to be between the 25th and the 50th percentile before stage-III operation. After a median period of 10 months, total cavopulmonary anastomosis (stage-III) has been performed in 13 patients and all survived.

died due to pneumococcal meningitis. The remaining 12 children are doing well and none has signs of heart failure. A hearing aid and medical therapy against seizures was necessary in 1 patient each. Two children received a pacemaker (AAIR) because of sinus node dysfunction. At present all survivors are in function class I or I–II of the NYHA classification.

3.1.8. Follow-up after complete Fontan

Follow-up was 7 months to 7 years. The average oxygen saturation was 95 ± 2% at last examination. A restrictive atrioseptectomy had to be reopened surgically in two children 10 and 15 months after stage-III respectively. Both survived and recovered completely with normal sinus rhythm. At the age of 2.5 years one of the 13 survivors

4. Discussion

The prevalence of HLHS is 0.16–0.26 of 1000 live births [5,6]. With about 750 000 live births in Germany the incidence of HLHS can be estimated as about 120 to 200 per year. Adding the other European countries it becomes obvious that there is severe shortage of donor hearts for that age group in Europe. In addition, patients after cardiac transplantation need long-term immunosuppression with its well known risks of infections, development of malignancies, secondary organ damage, accelerated coronaropathy, and limited cardiac survival [7]. Therefore we decided in 1989 to offer the staged reconstruction in parallel with cardiac transplantation for patients with HLHS or similar complex lesions. This report presents the results of the staged reconstructive procedure performed at our institution from the beginning to June 1998.

The acquired experience over time is mirrored by the improved results for typical HLHS which are documented by actuarial survival curves of three consecutive time periods. The hospital survival rate improved from 60% during the first time period to about 90% during the last time period.
A successful bidirectional cavopulmonary anastomosis leads to significant reduction of ventricular and pulmonary volume load [13,14] resulting in an improved survival rate after the final Fontan operation [15,16]. All patients of the present study who had undergone stage-III operation survived. The 4 year actuarial survival has increased over time and it seems reasonable to suppose that it will be between about 60% and 75% for patients who had undergone stage-I procedure during the third time period. Follow-up after Fontan operation is 7 months to 7 years. There was one late death due to meningitis but the remaining 12 patients are doing well and none has signs of heart failure. Recently it has been shown that several years after the Fontan operation there seems to be no difference in exercise performance between patients with a single right ventricle, who had had an initial Norwood procedure, and those patients with a single left ventricle, who did not have an initial Norwood operation [17].

The decision to change the three newborns with aortic atresia, hypoplastic ascending aorta, and interrupted aortic arch but two ventricles into a single ventricle physiology was governed by the concern about the long-term consequences. The reported biventricular procedure [18] used one valved conduit from the left ventricular apex to the descending aorta and a second conduit between the ascending and the descending aorta. Besides the disturbed left ventricular integrity, both conduits will not grow. Therefore the classical Norwood procedure was preferred since it allows growth of the aortic arch.

5. Conclusions

The presented data suggest that the staged reconstructive procedure is a realistic option for many of the newborns with typical HLHS and similar complex lesions regardless of the morphologic subtype or the diminutiveness of the ascending aorta. The reconstructions lead to an acceptable quality of life. A stage-I procedure performed during the first week of life will anticipate the development of an excessive pulmonary blood flow. By this means both unnecessary ventricular dilatation and the disadvantages of a foramen ovale, which may become progressively restrictive, are avoided at the same time. Preoperative atrioseptostomia seems to be unnessary. However, newborns with HLHS presenting a
severely dilated right ventricle and impaired ventricular function should, if at all, be candidates for transplantation rather than for reconstructive procedures.

References


Appendix. Conference discussion

Dr V. Hraska (Bratislava, Slovakia): Our experience with the treatment of hypoplastic left heart syndrome is limited. One and a half years ago we introduced this program in our institution; and up until now 15 patients have undergone the treatment of hypoplastic left heart syndrome, with 13 survivors, so we lost 2 patients. My question concerns risk predictors for bad outcome. Do you not think that with the proper preoperative care you can at least partially eliminate these risk factors?

Dr Breymann: We fully agree with that. And we have learned that we have to perform the Norwood procedure earlier than we have done in the first time frame. As you remember, we showed that the median time to the Norwood procedure was about 15 days, but there were patients where we waited at least for 3 months. During the very first time frame we feared that the pulmonary vascular resistance could be too high, but this was disastrous. I agree fully, we have to have that in mind to get better results.

Dr A. Corno (Genolier, Switzerland): I did not understand your point regarding the obstructive patent foramen ovale. It was believed that in the first few days of life the presence of obstructive patent foramen ovale was useful in hypoplastic left heart to maintain adequate pulmonary hypertension and to provide better systemic perfusion retrograde via the patent ductus arteriosus. So to my knowledge the Rashkind procedure has never been performed in a hypoplastic left heart. Your experience is different. My question is, what do you do now once you have evidence of obstruction at the level of the patent foramen ovale? Do you anticipate surgery? Do you perform Rashkind? Or can you explain why you have seen this picture?

Dr Breymann: In general, it is the same question. We try to operate earlier, before the foramen ovale gets too restrictive. Our results are all retrospective. But we think that the impact of a high pulmonary vascular resistance is the reason for getting worse results with obstructive venous return. That is what we found. But I think it does not matter if we try to operate very early after birth. We believe that this is the main key. The ventricles got large by waiting for the procedure, and in our opinion that was one of the main reasons for getting bad results.

Dr B. Sethia (Birmingham, UK): I want to ask you a question about the right ventricular volumes that you measured and specifically in the postoperative period whether you have any information about the way those volumes change in relationship to the size of the shunt that you placed. Because our experience has been that a smaller size shunt, in the main, results in a better outcome. And I wonder if you have any data to correlate the postoperative appearances of the right ventricle and physiology of the right ventricle with outcome and shunt size?

Dr Breymann: All that I can say is that we have decided, empirically, that the infants weighing 3.4 kg or more receive a 4 mm PTFA shunt. Below that weight we are using a 3.5 mm PTFA shunt.

Dr Sethia: An interesting observation. Because we found that nearly all patients would benefit with a 3 mm shunt, and it is very rare that we would use a 3.5 mm and never a 4 mm shunt now. So it is slightly different experience.

Dr Breymann: Okay. But I think we all have to learn a lot in this field.