Optimal conduit size for extracardiac Fontan operation☆

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Abstract

Background: Lack of conduit growth potential and thrombogenicity are the main drawbacks of the extracardiac Fontan operation (ECFO). Optimal size of the conduit according to the patients age and inferior vena cava diameter has not been established. Objectives: We set out to ascertain whether the optimal dimensions of the conduit could be determined before an ECFO. Methods: Actual and expected age-related inferior vena cava diameters were compared with the extracardiac conduit diameter in 20 patients after ECFO. In 50 other pediatric and adult patients, the distance between intrapericardial part of the inferior vena cava and the undersurface of the right pulmonary artery (IVC–RPA) was measured. Cases of conduit thrombosis were analyzed. Results: The actual diameter of the inferior vena cava was variable and has a weak correlation with anthropometric data and expected diameter (R = 0.07 ± 0.23, P = 0.32 ± 0.76). The IVC–RPA distance correlated with height (R = 0.87, P = 0.0001), but was also variable. At the age of 2–4 years and body weight 12–15 kg IVC diameter and IVC–RPA distance are equal to 60–80% of adult values. Conduit thrombosis developed in two patients with unfavorable Fontan hemodynamics and oversized conduits. Conclusions: Considering the inferior vena cava size, ECFO may be performed at the age of 2–3 years and at a body weight 12–15 kg, when a hemodynamically optimal almost adult sized conduit can be implanted. Optimization of the conduit is necessary on the basis of the actual inferior vena cava diameter and IVC–RPA distance. Anticoagulation postoperatively should be considered to prevent conduit thrombosis in patients with suboptimal Fontan circulation © 2000 Elsevier Science B.V. All rights reserved.

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1. Introduction

The extracardiac Fontan operation (ECFO) operation is thought to bear some theoretical advantages over other types of Fontan connections, such as avoidance or minimization of pre-pulmonary stenosis, avoidance of extensive atrial suture lines, cardioplegia and hypothermia, as well as optimized laminar flow in the conduit and a lower frequency of arrhythmias [1–5]. Whether ECFO has real advantages in the long term still has to be established.

Lack of growth potential and thrombogenicity of the conduit are the major drawbacks and main points of criticism of the ECFO [6]. In the literature there are no data on the optimal size of the conduit in accordance with the patients anthropometric data, diameter of the inferior vena cava (IVC) and distance between the cardiac segment of the IVC and the lower surface of the right pulmonary artery (IVC–RPA), where the conduit is usually implanted during ECFO.

Conduit performance seems to be an important issue of the ECFO. Therefore, the purpose of this study was to find out whether optimal dimensions of the conduit could be established before ECFO.

2. Patients and methods

Thirty-six patients underwent ECFO in our institution. Twenty of them had cardiac catheterization and angiography before and after surgery and were studied retrospectively. The ages of the patients ranged from 1.3 to 27.4 years (median 3 years) and body weight from 8.3 to 54.5 kg (median 12.8 kg). The anatomical diagnoses are presented in Table 1. Azygous continuation of the inferior vena cava was observed in one patient. Cavopulmonary shunt was performed previously in 16 patients.

Intraoperatively the diameter of the inferior vena cava was measured after dissection of its pericardial attachment.
During ECFO Gore–Tex® e-PTFE conduit (W.L. Gore & Assoc., Elkton, MD) of maximal diameter was implanted. For most children the conduit was 20–22 mm in diameter and 24 mm in adults. A Gore–Tex® conduit of 16 mm was implanted in one patient; 18 mm in three; 20 mm in eight; 22 mm in five and 24 mm in three patients (two adults). Conduit length was recorded intraoperatively in 19 of 20 patients. It was 3.0–3.5 cm in eight patients; 4.0–4.5 cm in nine; and 5.0 and 6 cm in two patients. The length of the conduit was 1–2 cm shorter than the intraoperatively measured IVC–RPA distance because the anastomosis to the IVC always included a short rim of right atrial tissue to create an anastomosis of conduit diameter. Anastomoses with the undersurface of the right pulmonary artery was performed in an oblique fashion to enlarge the pulmonary bifurcation and to obtain a larger anastomoses than the conduit diameter. Fenestration was performed in 12 patients and was found during follow-up to have closed spontaneously in nine patients. In one patient fenestration was closed with an umbrella device during control cardiac catheterisation. Maximal angiographic diameter at the cardiac end of the IVC at its entrance to the pericardial cavity was measured pre- and postoperatively [7], as well as the diameter of the conduit and the conduit distal and proximal to the anastomoses. There were no instances of anastomosis stenosis or conduit external shape changes. Actual age, height, body surface area and weight-related and expected calculated [8] IVC diameter were compared with the extracardiac conduit diameter. The expected age, height, weight and body surface area that related as the inferior vena cava diameter was calculated according to Steinberg et al. [8].

To control conduit patency and thrombosis, all of the patients underwent follow-up echocardiographic control angiography 1–30 months (median 8 months) after surgery and eight patients also underwent magnetic resonance imaging (MRI). In all cases the smallest diameter of conduit and conduit proximal and distal anastomoses were measured.

In 50 other non Fontan patients aged between the first day of life and 77 years (median 13 years), body weight from 2.4 to 118 kg (median 43.5 kg) and body surface area between 0.18 and 2.19 m² (median 1.33 m²) with congenital and coronary heart disease without pulmonary artery hypoplasia, the distance between the cardiac segment of the IVC, just above the diaphragm and the undersurface of the right pulmonary artery, was measured after median sternotomy (31 patients) and during cardiac catheterization (19 patients).

No postoperative anticoagulation treatment except aspirin was used in the studied patients.

2.1. Statistical analysis

Statistical analysis was performed using the linear regression and Spearman rank correlation using SPSS for Windows 8.0. The difference was considered statistically significant when the P-value was ≤0.05.

3. Results

3.1. Diameter of the inferior vena cava

Actual diameter of the IVC in 20 patients varied between 7.7 and 27.6 mm and has a weak correlation with age (R = 0.07, P = 0.76, Fig. 1), weight (R = 0.23, P = 0.33), height (R = 0.19, P = 0.43) and body surface area (R = 0.17, P = 0.47). Normal age related expected diameter varied from 13.2 to 20 mm. The relationship between actual and expected diameter of the IVC revealed a weak correlation (R = 0.07, P = 0.76, Fig. 1). At age 2–4 years mean actual diameter of the IVC was 16.6 mm, which is 81% of mean normal adult diameter (20.4 mm) [7,9].

3.2. IVC–RPA distance

Intraoperative and angiographic measurements in our patients revealed that the IVC–RPA distance varied from 2.3–8.0 cm. At ages less than 2 years (N = 12) it was equal

![Fig. 1. Actual (closed circles) and age-related calculated (open circles) inferior vena cava diameter in 20 patients after ECFO. Actual IVC diameter is variable and does not correlate with the calculated diameter (R = 0.07, P = 0.76). Note variations of the IVC diameter in patients of the same age. Squares represent markedly (more than 1.5 times over the actual IVC diameter) oversized conduits. See text for details.](image)
to 3.5 cm (2.3–3.9 cm), at ages 2–4 years (N = 9) it was 4 cm (3–5.1 cm), at ages 10–16 years (N = 7) it was 5.7 cm (4.6–7.0 cm), and in adults (N = 22) 6.5 cm (4.5–8.0 cm). The most pronounced correlation was found between height and IVC–RPA distance (R = 0.87, N = 50, P < 0.0001, Fig. 2) and IVC–RPA distance and body surface area (R = 0.84, P < 0.0001). The correlation with age and weight was less pronounced (R = 0.67 and R = 0.78 respectively, P < 0.0001). At ages 2 to 4 years this distance approached 60% of that found in adults.

3.3. Conduit diameter

Conduit to actual IVC diameter ratio was 0.66–2.0 (median 1.33, Fig. 3). Conduit to expected IVC diameter ratio was 1.0–1.67 (median 1.38, Fig. 3). There was no correlation between real conduit to IVC ratio and expected age-related diameter ratio (R = 0.09, P = 0.72, Fig. 3).

Analysis of the postoperative angiograms of the 20 patients after ECFO revealed that in six patients an oversized (more than 1.5 times) conduit had been implanted in relation to the actual IVC diameter (Fig. 3).

3.4. Conduit thrombosis

We observed two cases of partial conduit thrombosis: one in a 3-year-old child with Ebstein anomaly, who underwent closure of the tricuspid valve as a newborn, together with an atrioseptectomy and central aorta-pulmonary shunt. She underwent ECFO at 3 years of age with a 20 mm conduit, which retrospectively was markedly oversized (2.0 times greater than the actual IVC diameter, Fig. 4A). Several months later she developed congestive heart failure and protein-losing enteropathy. She underwent surgery at 4 years of age because of partial thrombosis of the conduit detected by angiography and MRI. Conduit replacement with a 16 mm aortic homograft was performed. Intraopera-

tively partial thrombosis of the conduit was found (Fig. 5). She is now free of symptoms. Another case of conduit partial thrombosis was observed in a 27-year-old patient with an interrupted inferior vena cava and an ayzygos continuation, who underwent surgery early in our series of ECFO, in whom only a short segment of the IVC, where the hepatic veins drained, was connected with a 24 mm Gore-Tex® tube to the right pulmonary artery (conduit to IVC ratio was 1.9), and the main Fontan pathway was the left-sided ayzygos vein. Postoperatively the patient received no anticoagulation treatment except aspirin, but when control cardiac angiography and MRI revealed partial conduit thrombosis he was placed on an anticoagulation regimen with warfarin to target the International Normalized Ratio (INR) of 2–3.

In four other patients with oversized conduits (conduit to IVC diameter ratio 1.5–1.7) no conduit thrombosis was observed during follow-up echocardiographic and angiographic control studies.

4. Discussion

The ECFO was performed with increased frequency in patients with a single ventricle physiology [5]. Proponents of the ECFO postulated that theoretically it may decrease the frequency of postoperative myocardial dysfunction and atrial arrhythmias, improve the hydrodynamics of the Fontan connection, decrease the incidence pre-pulmonary stenosis and fenestration and as a result improve the long-term prognosis [2,4,5,10–12]. The most controversial issue of the ECFO is the absence of growth potential of the implanted conduit (usually Gore–Tex®), which may necessitate conduit replacement in the growing child and increase the risk of partial conduit thrombosis and the need for long-term anticoagulation treatment [6].

The normal adult IVC expiratory mean diameter is 20.4 mm with variations that range from 10 to 36 mm [7,9,13–17] without a strong correlation with age, sex, height,
weight or corporeal area. In children, such a correlation exists up to 14–15 years of age [8]. However, even a 2-year-old child may have an adult sized IVC or the converse (Fig. 4A,B) [7,8]. The diameter of the IVC is increased in patients with congenital and acquired heart diseases, particularly in the presence of heart failure with increased right atrial pressure [7].

After lateral tunnel Fontan operation, the atriopulmonary anastomosis is often obstructive when compared with some of the caval areas (Fontan area). Therefore, the diameter of Gore-Tex tube in children should be equal or somewhat larger than the diameter of the cardiac end of the IVC in order to neutralize the absence of growth potential of the conduit [4]. However, optimal diameter of the conduit has to be determined, despite the fact that ECFO appears to solve the problem of pre-pulmonary stenosis.

The upper limit of critical conduit to IVC diameter ratio is approximately 1.5, without a substantially negative hydrodynamic consequence [5]. In 14 of 20 of our patients the conduit used oversized the IVC diameter by 1.2–1.5 times (Fig. 3), which is thought to be optimal for hemodynamic performance [5]. Such moderate oversizing can be ignored with respect to the decreased risk of the necessity for conduit replacement in the future but individual variations of the IVC diameter in children and adults prevent the prediction of which size of conduit will become inadequate with a child’s growth. Conduits implanted without knowing the actual IVC diameter may result in a discrepancy between conduit to IVC diameter and suboptimal Fontan circulation.

There are two directional growth potential restrictions of the implanted prosthesis: diameter and length. It is doubtless that conduit growth cannot be expected, but it is also doubtful whether implantation of the maximum possible sized conduit is necessary, even in some adults, because of the variable diameter of the IVC.

The optimum conduit length for ECFO also has not yet been established. In a reported large series of ECFO, the lengths of the conduits were not given [2,10,12,18]. There is no data in the anatomical and surgical literature on the IVC–RPA distance. Intraoperative measurement of this distance permits the use of a shorter conduit, because the anastomosis of the conduit with the IVC always includes a short rim of the right atrial wall, and thus has longitudinal and transverse growth potential. Thin (usually less than 1000 \( \mu \text{m} \)) neointimal ingrowth occurs in the e-PTFE graft [19] and cannot influence conduit performance. Theoretically a conduit that is too short may become the cause of distortion of the right pulmonary artery and compression of the pulmonary veins as the patient grows, but we and others have not observed such complications after ECFO [2,3,10,11,18]. Perhaps the optimal age for ECFO is 2–4 years, when the mean diameter of the cardiac end of the IVC and RPA–IVC distance approaches 60–80% of the adult size. The assessment of the actual IVC diameter before ECFO appears to be important so that the implantation of a hemodynamically optimal conduit without significant oversizing can take place. Even conduit of 20 mm diameter can be optimal or oversized in a small child (Fig. 4A,B).

We now prefer implantation of the extracardiac conduits which oversize the diameter of the IVC by no more than 20%. If a patient needs the conduit to be replaced later, this seems to be technically easy and can be performed with partial bypass [20].

Fig. 4. Angiograms of the IVC and conduits of two patients with similar anthropometric data. In both patients 20 mm conduit prosthesis was implanted during ECFO. (A) Two-year-old patient with weight 12 kg, height 72 cm and body surface area 0.5 \( \text{m}^2 \). Conduit to inferior vena cava diameter ratio is 2.0. (B) Two-year-old patient with weight of 13 kg, height 91 cm and body surface area 0.7 \( \text{m}^2 \). Conduit to IVC ratio is 1. There was a pronounced difference in IVC diameter between the two patients.
Thromboembolic complications are one of the major concerns after all types of Fontan operation, and occur in as many as 20% of cases [21–23]. Use of anticoagulation treatment is discussed controversially after Fontan operation [21]. After Fontan operation some groups recommend anticoagulation with Warfarin for at least 12 months, followed by a life-long aspirin regimen [3,11,20,23].

Neointima, which develops in the Gore-Tex prosthesis 3–6 months after experimental replacement of caval veins, is thicker at the anastomosis site and thinner in the middle part (700–1000 μm) where sometimes thrombotic focuses develop [19].

In two of six of our patients with considerably oversized conduits, partial thrombosis developed during the first year after surgery. When the left-sided azygous vein bears the main Fontan flow, the conduit size should be adjusted to the size of the IVC entrance into the right atrium to minimize turbulent flow and the risk of thrombosis, which developed in one patient with such an anomaly. Therefore, anticoagulation treatment should be considered in patients with suboptimal Fontan circulation in whom a Gore-Tex conduit is implanted.

In conclusion, when considering the inferior vena cava size, ECFO in children may be performed optimally at 2–4 years of age and at a body weight of 12–15 kg when the diameter of the inferior vena cava and the distance between the IVC and the right pulmonary artery approaches 60–80% of the adult value and a conduit can be implanted without significant oversizing and negative hemodynamic consequences. Optimization of the conduit dimensions is necessary in relation to the pre- or intraoperatively measured actual IVC diameter and the IVC–RPA distance. When an e-PTFE graft conduit is used, considering the absence of true endothelialization of the prosthesis, the use of long-term anticoagulation for at least 6 months after the operation is important to prevent conduit thrombosis, which may develop as early as during first year after surgery, especially in the presence a suboptimal Fontan circulation.

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References


