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Influence of Pulmonary Artery Size on Early Outcome After the Fontan Operation

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Background. Small pulmonary arteries (PAs) are often considered as a contraindication for the Fontan operation (FO). The aim of this study was to evaluate if the PA size is still one of the major impact factors on the postoperative early outcome.

Methods. Data of 146 patients, with a median age of 2.0 years (range, 1.5 to 18 years) and a median weight of 12.45 kg (range, 7.7 to 64.7 kg) who underwent a modified FO in the same center between 2007 and 2012 were retrospectively analyzed with respect to the traditional McGoon ratio, Nakata index, and modified indices (measuring the narrowest diameters).

Results. Patients with a McGoon ratio of 1.6 or less (modified ≤ 1.2) or a Nakata index of 150 mm²/m² or less (modified ≤ 100 mm²/m²) were not at a higher risk of longer mechanical ventilation ($p = 0.87$ [0.1] and $p = 0.68$ [0.52], respectively), longer stay ($p = 0.52$ [0.18] and

$p = 0.54$ [0.38], respectively) in the intensive care unit, prolonged hospital stay ($p = 0.08$ [0.26] and $p = 0.22$ [0.29], respectively) or effusions ($p = 0.25$ [0.37] and $p = 0.13$ [0.06]), respectively). Younger and smaller children tended to have smaller PAs, but younger age (<24 months) and lower weight (<12 kg) were not predictive for poor early postoperative outcome.

Conclusions. Small PAs do not significantly affect the early postoperative period after FO. In our opinion, there is no need to postpone the FO due to "smaller" PAs. The palliative procedures performed before FO to increase the size of the PA at the expense of volume overload of the single ventricle and the possible complications of prolonged cyanosis must be carefully weighed.

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In 1977 Choussat and colleagues [1] presented 10 criteria for a reasonable patient selection before establishing a univentricular circuit. These "10 commandments" were age older than 4 years, adequate ventricular function, a competent atrioventricular valve, normal systemic venous drainage, normal right atrial volume, sinus rhythm, adequate pulmonary artery (PA) size, absence of PA distortion, low PA pressure (PAP), and low pulmonary vascular resistance (PVR) [1]. During the last 3 decades, modifications of the initial procedure, such as the establishment of an extracardiac conduit or an intracardiac lateral tunnel, as well as application of a fenestration, drew attention to other hemodynamic features, and the original selection criteria have been adapted.

A remaining factor of concern before the Fontan pathway is the status of the pulmonary vascular bed. Although high PVR as a predictor for a negative outcome meets broad agreement [2, 3], the effect of the PA size is still controversial. Preservation of the single ventricle's function by volume reduction, on the one hand, and stimulating pulmonary artery growth via increased pulmonary blood flow, on the other hand, needs delicate hemodynamic balancing before the Fontan operation (FO) [4]. This study aims to evaluate the influence of preoperative PA size on early outcome after the univentricular palliation in a time of advanced interventional and operative procedures more than 40 years after Fontan's pathbreaking trial.

Patients and Methods

Between 2007 and 2012, 153 children underwent an intracardiac or extracardiac FO at the University Hospital in Munich. The study was approved by the Institutional Review Board of our institution. Informed consent for

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Abbreviations and Acronyms

AV	= Atrioventricular
DILV	= double-inlet left ventricle
FO	= Fontan operation
HLHS	= hypoplastic left heart syndrome
HRHS	= hypoplastic right heart syndrome
ICU	= intensive care unit
LAP	= left atrial pressure
LPA	= left pulmonary artery
PA	= pulmonary artery
PAP	= pulmonary artery pressure
PVR	= pulmonary vascular resistance
Qp:Qs	= ratio of pulmonary blood flow to systemic blood flow
RPA	= right pulmonary artery
SaO ₂	= arterial oxygen saturation
SD	= standard deviation
SV	= single ventricle
SVEDP	= systemic ventricular end-diastolic pressure
TGA	= transposition of the great arteries

treatment was obtained from the parents of each child. Retrospectively, their data, including age, weight, and height at FO were collected. Exact diagnoses of their univentricular circulation and the performed method of palliation (intracardiac tunnel with fenestration or extracardiac conduit) were registered. Furthermore, the early postoperative outcome for duration of mechanical ventilation, length of intensive care unit (ICU) and hospital stay, and duration of pleural or peritoneal effusions was evaluated. Our strategy was to leave the drainage tube in place until daily fluid loss was less than approximately 3 mL/kg.

Cardiac catheterization data before the FO were available in 146 patients. Preoperative hemodynamic data and possible interventional procedures were noticed. With help of an Xcelera Workstation (Koninklijke Philips N.V., Amsterdam, Netherlands) traditional McGoon ratio and Nakata indices were calculated in angiographic loops. All measurements were done twice by 2 independent examiners. The McGoon ratio includes the combined diameters of both PAs and the diameter of the aorta just above diaphragm, whereas the Nakata index comprises the cross-sectional area of both PAs (mm²) and the patient's body surface area (mm²) [5, 6]. Modified indices were also calculated using the narrowest diameters of both left and right PA, and only indexed measures of the PAs were statistically analyzed.

To evaluate the influence of PA size on the early postoperative outcome, the patients was divided into groups. The first criterion of division was a McGoon ratio of 1.6 or less vs more than 1.6, and the second was a Nakata index of 150 mm²/m² or less vs more than 150 mm²/m². Afterward, we took a closer look at the patients with very small PAs and compared those with McGoon modified ratios of 1.2 or less vs patients with ratios exceeding 1.2 and modified Nakata indices of 100 mm²/m² or less vs those exceeding 100 mm²/m². Cutoff points for grouping were

selected according to literature addressing the same or a similar discussion [7–10].

To evaluate if patients with prolonged effusions and longer hospital stay had significantly smaller PAs, we compared those with a hospital stay exceeding 14 days vs 14 days or less and patients with drainage exceeding 14 days with those who had an uncomplicated clinical course.

Further investigations focused on the influence of preoperative interventional procedures such as balloon dilatation or stenting of the PAs. Finally, weight and age within our cohort were analyzed with respect to their predictive value on the early postoperative outcome after the FO.

Statistical Analysis

Statistical analysis was performed with SigmaPlot 10 software (Systat Software Inc, Chicago, IL). Continuous variables are expressed as mean $\pm$ standard deviation or median (range). The 2-tailed Student *t* test or Mann-Whitney *U* test was performed for comparison of continuous variables between the groups and the Fisher exact test for categorical data. The Spearman rank correlation coefficient was used to evaluate the dependence between two nonparametric variables. A *p* value of less than 0.05 was considered significant.

Results

Demographic Data

In our study cohort, mean age at the FO was 3.6 ± 2.4 years (range, 16 months to 18 years), and mean weight was 14.3 ± 6.9 kg (range, 7.7 to 64.7 kg). The mean body surface area was 0.59 ± 0.2 m² (range, 0.39 to 1.65 m²). Most patients were diagnosed as having a hypoplastic left heart syndrome (*n* = 80). The detailed diagnosis of all children is presented in Table 1. In 56 patients (38.4%), an intracardiac lateral atrial tunnel with a 2- to 3-mm fenestration was established using a polytetrafluoroethylene patch, whereas 90 children (61.6%) underwent an extracardiac repair with a polytetrafluoroethylene conduit 18 mm in diameter (Gore-Tex Vascular Graft; W.L. Gore & Associates, Dundee, Scotland, United Kingdom). In only 2 children from the extracardiac conduit group, a 2- to 4-mm fenestration was made because of an increased PAP during the cardiac

Table 1. Cardiac Morphology

Variable	No. (%) (N = 146)
HLHS	80 (54.8)
HLHS variants	21 (14.4)
HRHS	20 (13.7)
DILV $\pm$ TGA	20 (13.7)
Others	5 (3.4)

DILV = double inlet left ventricle; HLHS = hypoplastic left heart syndrome; HRHS = hypoplastic right heart syndrome; TGA = transposition of the great arteries.

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