Somatic development long after the Fontan operation: Factors influencing catch-up growth

Masamichi Ono, MD,[a] Dietmar Boethig, MD,[b] Heidi Goerler, MD,[a] Melanie Lange, MS,[a] Mechthild Westhoff-Bleck, MD,[c] and Thomas Breymann, MD[a]

Supplemental material is available online.

Objective: As mortality and morbidity after the Fontan operation has improved, long-term outcome, including developmental aspects, have become more important. To understand the long-term effects of this operation, we followed somatic development for up to 15 years.

Methods: We evaluated 90 patients who underwent the Fontan operation between 1984 and 2004 (mean follow-up, 11.8 ± 4.2 years). The modified Fontan operations were atriopulmonary anastomosis (n = 19) and total cavopulmonary connection (n = 71). Mean age at the time of surgical intervention was 5.5 ± 4.8 years. Weight, height, and body mass index were evaluated preoperatively and postoperatively and given as percentiles on a normal growth curve.

Results: Postoperative weight, height, and body mass index reached the 47.2 ± 35.6, 37.9 ± 30.4, and 41.6 ± 31.2 percentiles, which were significantly better than preoperative values (the 21.6 ± 25.9, 25.9 ± 25.7, and 20.0 ± 25.1 percentiles). Although neither early surgical intervention nor anatomic features affected postoperative growth, early Fontan completion demonstrated better somatic development in subgroups of tricuspid atresia. Prior bidirectional Glenn shunting provided better weight gain before the Fontan operation. Prior atrioseptectomy, central shunt, and pulmonary artery reconstruction were associated with impaired somatic development. Reoperation and catheter-based intervention improved somatic development.

Conclusions: Long-term catch-up growth can be observed in patients after the Fontan operation. Early volume-unloading procedures might lead to better somatic growth. Prior atrioseptectomy, central shunt, and pulmonary artery reconstruction are associated with impaired weight and height gain, implying that the severity of the underlying diseases affects postoperative somatic development.

From the Divisions of Thoracic and Cardiovascular Surgery,[a] Pediatric Cardiology and Intensive Care Medicine,[b] and Cardiology and Angiology,[c] Hannover Medical School, Hannover, Germany.

Supported in part by grants from The Uehara Memorial Foundation and The Mochida Memorial Foundation for Medical and Pharmaceutical Research.

Received for publication Jan 13, 2007; revisions received July 14, 2007; accepted for publication Aug 2, 2007.

Address for reprints: Masamichi Ono, MD, Division of Thoracic and Cardiovascular Surgery, Hannover Medical School, Carl-Neuberg-Str 1, 30625 Hannover, Germany. (E-mail: Ono.Masamichi@MH-Hannover.DE).

J Thorac Cardiovasc Surg 2007;134:1199-206

0022-5223/$32.00

doi:10.1016/j.jtcvs.2007.08.002

The Fontan operation was first performed in patients with tricuspid atresia as a functional repair.[1] Modifications of the Fontan operation,[2] staging with a bidirectional cavopulmonary shunt (ie, the bidirectional Glenn [BDG] shunt),[3] and creation of fenestration in the Fontan pathway[4] have reduced the mortality and morbidity associated with this operation. Today, the Fontan operation is used to treat a wide range of congenital heart defects with functional single ventricles. Improvements in preoperative management, surgical techniques, and intensive care medicine have made the Fontan operation a safe procedure at younger ages than ever before, and many reports describe excellent results from the Fontan operation in small children.[5-7] The early elimination of cyanosis and volume overload in the functional single ventricle seems to be beneficial, but whether these modifications provide better postoperative somatic development is unknown. Despite the improvements in perioperative mortality, few studies have assessed the potential for postoperative growth in children with Fontan circulation.

Abbreviations and Acronyms
APA = atriopulmonary anastomosis
BDG = bidirectional Glenn
BMI = body mass index
CHD = congenital heart disease
PLE = protein-losing enteropathy
TCPC = total cavopulmonary connection

In the present study we evaluated somatic development over the long-term after the Fontan operation and investigated factors influencing somatic development. Moreover, the benefit of surgical and catheter-based interventions after the Fontan operation was also evaluated.

Materials and Methods

Patients

Between 1984 and 2004, 121 consecutive patients with functionally univentricular hearts underwent Fontan-type operations at our institution. There were 10 early deaths and 6 late failures (death or heart transplantation). Among the 105 survivors with Fontan circulation, 90 were followed up by serial measurements of weight and height. Fifteen patients are known to be alive with a Fontan circulation but could not be traced by means of regular follow-up. The characteristics of 90 patients are shown in Table 1.

The mean age at the time of the Fontan operation was 5.5 ± 4.8 years, ranging from 0.6 to 20 years. The mean follow-up period was 11.8 ± 4.2 years. Modifications of the Fontan operation included atriopulmonary anastomosis (APA; n = 19), performed until 1989, and total cavopulmonary connection (TCPC; n = 71), performed from 1988. In patients treated with TCPC, a fenestration was created at the time of Fontan completion in 18 patients, and a BDG shunt was used before the Fontan operation in 20 patients (Table 1).

Data Collection of Somatic Development

Somatic development was evaluated before and 1, 2, 5, 7, 10, 12, and 15 years after the Fontan operation by using body weight and standing height, which were related to the standard growth percentiles (normal is 50 percentile), as described previously.[8,9] The body mass index (BMI) was calculated and related to the standard growth percentiles. For 20 patients who underwent BDG shunting, somatic development was also evaluated before and after BDG shunting. Data were collected from the patients' records when they were available at the Hannover Medical School. For patients followed up elsewhere, data were collected by means of fax transmission.

Informed consent was obtained from all patients, their parents, or both. The Ethics Committee of the Hannover Medical School approved the study protocol.

Statistical Analysis

Values are expressed as the mean ± standard deviation. Data were analyzed by using the SPSS statistical software system (SPSS, Inc, Chicago, Ill). The primary outcome was determined as the weight, height, and BMI at the final follow-up examination. This outcome was then compared with preoperative data by using the paired Student *t* test. The primary outcome was analyzed with a variety of categoric variables by using the unpaired Student *t* test and with metric variables by using correlation analysis. Multivariate analysis was done by using a linear regression model to identify the risk factors that predict the primary outcome. Potential interactions among variables included in the model were analyzed by means of contingency table methods.

TABLE 1. Patient characteristics (n = 90)

Variable	Total (n = 90) No.	Percentage
Diagnosis		
Tricuspid atresia	19	21.1
Predominant right ventricle	30	33.3
Transposition of the great arteries	47	52.2
Heterotaxy	7	7.8
Atrioventricular valve anomaly	19	21.1
Systemic venous anomaly	8	8.9
Pulmonary venous anomaly	4	4.4
Palliative procedure		
Atrioseptectomy	13	14.4
Coarctation repair	8	8.9
Blalock–Taussig shunt	38	42.2
Central shunt	19	21.1
Pulmonary artery banding	10	11.1
Pulmonary artery reconstruction	12	13.3
Modification of Fontan completion		
Atriopulmonary anastomosis	19	21.1
Total cavopulmonary connection	71	78.9
With fenestration	18	20.0
After bidirectional Glenn shunt	20	22.2

Results

Postoperative Body Weight, Height, and BMI

The primary outcomes determined by weight, height, and BMI at the last follow-up were the 47.2 ± 35.6, 37.9 ± 30.4, and 41.6 ± 31.2 percentiles, respectively (Figure 1). These values were significantly higher than the patients' preoperative values, which were on the 21.6 ± 25.9, 25.9 ± 25.7, and 20.0 ± 25.1 percentiles ($P < .01$ in all variables). The number of patients smaller than the fifth percentile decreased from 34 (37.8%) to 13 (14.4%) in weight, from 27 (30.0%) to 12 (13.3%) in height, and from 36 (40.0%) to 9 (10.0%) in BMI, whereas the number of the patients larger than the 85th percentile increased from 4 (4.4%) to 17 (18.9%) in weight, from 3 (3.3%) to 9 (10.0%) in height, and from 4 (4.4%) to 11 (12.2%) in BMI. Body weight and BMI have significantly improved by 1 year after the operation. Height has significantly improved by 2 years postoperatively (Table E1).