Right Ventricular Outflow Tract Growth in Infants With Palliated Tetralogy of Fallot

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Background. **In symptomatic neonates with tetralogy of Fallot with pulmonary stenosis (TOF/PS), limited contemporary data exist regarding the effect of a modified Blalock-Taussig shunt (mBTS) on pulmonary valve (PV) and pulmonary artery (PA) growth and on the incidence of PV preservation at the time of complete repair.**

Methods. **We retrospectively studied patients who underwent repair of TOF/PS from 2000 to 2012 at our center. In neonates with TOF/PS palliated with an mBTS, we assessed PV annulus, main PA (MPA), and branch PAs (left PA [LPA] and right PA [RPA]) size by echocardiography before mBTS and surgical repair.**

Results. **Of 172 patients with TOF/PS, 40 (23%) were palliated with an mBTS at a median age of 23 days, and 31 met criteria for echocardiographic analysis. Palliated patients had hypoplastic PV, MPA, RPA, and LPA at baseline. All structures had significant absolute growth before surgical repair ($p < 0.001$). PV and MPA *Z* scores were unchanged, whereas branch PAs showed significant improvement (RPA, $p = 0.03$; LPA, $p = 0.008$). A PV-sparing repair was performed in 116 of 132 primary repairs (88%) and in 10 of 31 palliated patients (32%). At a median follow-up of 4.0 years (range, 0.7 to 12.6 years), no palliated patients required reintervention for right ventricular outflow tract obstruction.**

Conclusions. **Significant absolute growth of the PV, MPA, RPA, and LPA occurs after an mBTS in neonates with TOF/PS. Branch PA size normalizes before complete repair. In one-third of patients with hypoplastic PVs, absolute growth after an mBTS may have facilitated valve-sparing repair.**

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The favored approach to complete surgical repair of tetralogy of Fallot with pulmonary stenosis (TOF/PS) has varied over time. Initially, most repairs were performed via a right ventriculotomy with transannular patch (TAP) placement and a goal of minimizing residual right ventricular outflow tract obstruction, while accepting free pulmonary insufficiency as an outcome [1]. However, more recent data indicate that right ventricular dysfunction [2–4], poor functional status [5], and arrhythmias [6, 7] often develop in such patients, and they commonly undergo reoperation for pulmonary valve (PV) replacement [8]. Several surgical strategies have been used with the intent to preserve a functional PV, including commissurotomy, passage of rigid dilators, PV augmentation, and sequential intraoperative balloon pulmonary valvuloplasty [9, 10]. Nonetheless, TAP repair remains the most common approach to repair of TOF/PS in the modern era [11].

Most patients with TOF/PS now undergo primary complete repair in the first year of life, most commonly between age 3 and 6 months [11]. In symptomatic neonates (aged <30 days) with an unacceptable degree of cyanosis, some centers advocate for primary repair in the neonatal period, citing comparable mortality rates and elimination of shunt risks such as thrombosis and pulmonary artery (PA) distortion [12, 13]. Variable rates of a TAP approach to relieve right ventricular outflow tract obstruction are reported in young infants, ranging from 24% to 84% [9, 11, 13, 14]. Patients with a significantly hypoplastic PV who undergo a neonatal repair are more likely to have a TAP, with some institutions using empiric TAP repairs in neonates with severe PV hypoplasia [9, 12, 14].

Alternatively, symptomatic neonates with TOF/PS may undergo initial palliation with a modified Blalock-Taussig shunt (mBTS) before complete surgical repair. Advocates of this approach suggest that initial palliation may optimize the chances for preservation of a functional PV at the time of the later repair [10, 15]. Data from The Society of Thoracic Surgeons Congenital Heart Surgery Database indicate that initial surgical intervention for TOF/PS during the neonatal period is fairly equally divided between palliation and primary repair [11].

The effect of initial palliation with an mBTS on PV growth, PV preservation at the time of repair, and the durability of PV-sparing (PVS) repairs remains unclear. Available data regarding growth of the PV and PAs in

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palliated patients with TOF/PS are derived from postmortem analysis, cardiac catheterization, and intraoperative measurements [16–19]. Importantly, these studies were conducted in an earlier era, when initial palliation was rarely performed in the neonatal period, and complete repair was typically deferred for several years, resulting in a protracted interstage period.

This study examined longitudinal growth of the PV annulus and the main (MPA) and branch PAs (left [LPA] and right [RPA]) by echocardiography in infants with TOF/PS who were palliated with an mBTS in the neonatal period. We hypothesized that palliation with an mBTS in patients with TOF/PS would not adversely affect the growth of the PV annulus and PAs.

Material and Methods

This single-center, retrospective study was approved by Ann & Robert H. Lurie Children's Hospital of Chicago Office of Research Integrity and Compliance. The need for written informed consent was waived.

Study Patients

All patients with TOF/PS, including those with double-outlet right ventricle of tetralogy type, who underwent surgical repair at Ann & Robert H. Lurie Children's Hospital of Chicago between January 2000 and September 2012 were identified from the cardiac surgical database for study inclusion. Excluded were patients with TOF with pulmonary atresia, absent PV, atrioventricular septal defect, or other complex congenital heart disease. Also excluded were patients who underwent surgical or percutaneous intervention of the right ventricular outflow tract between birth and complete surgical repair.

Data Collection

Demographic data included gender, age, and weight at palliation with an mBTS, if applicable, and at surgical repair. Operative reports were reviewed to document the surgical approach to right ventricular outflow tract obstruction for all patients (TAP, PVS repair, or right ventricle-to-PA conduit). For patients palliated with an mBTS, the PV morphology by direct intraoperative observation, the timing of and indication for any percutaneous or surgical right ventricular outflow tract reintervention, and the operative or postoperative deaths were recorded.

Growth of the PV and PAs

For each patient with available echocardiographic images, we selected 3 studies for review: the initial postnatal study, the study before the mBTS, and the study before the surgical repair. In some patients, a single echocardiogram was performed between birth and mBTS placement. One observer measured the absolute diameters of the PV annulus, MPA, RPA, and LPA. A second independent observer measured the same parameters for one-third of the patients. The physician–readers obtained measurements in accordance with the American Society of Echocardiography guidelines, and Z scores were generated [20]. The PV was measured during systole in the parasternal long-axis or short-axis view, whichever view best demonstrated the valve hinge points. The MPA, LPAs, and RPAs were measured during systole in the parasternal short-axis view.

Statistical Analysis

Baseline characteristics were summarized using conventional descriptive statistics. The general linear model repeated-measures of analysis of variance with a Greenhouse-Geisser correction was used to assess for significant change in the PV, MPA, RPA, and LPA absolute diameters and Z scores from the time of the initial postnatal echocardiogram to the time of surgical repair. Interreader reliability was assessed by interclass correlation. SPSS 21 statistical software (IBM Corp, Armonk, NY) was used for all analyses.

Results

Between January 2000 and September 2012, 172 patients underwent repair of TOF/PS, 40 (23%) of whom were palliated with an mBTS before surgical repair. Complete echocardiographic data were available for 33 of 40 palliated patients. The 7 patients with incomplete echocardiography data were excluded from further analysis, as were 2 patients who underwent right ventricular outflow tract intervention before complete surgical repair (Fig 1).

Demographics and selected surgical characteristics of the study patients are found in Table 1. The median age at mBTS was 23 days (range, 2 to 212 days). Only 4 patients underwent palliation at an age older than 60 days (range, 79 to 212 days), 3 of which were performed as emergencies. One patient had severe PA hypoplasia, the second had a history of prematurity, chronic lung disease, and severe respiratory syncytial virus bronchiolitis, and the third had a history of severe aspiration pneumonia, subglottic stenosis, and urgent tracheostomy. One patient scheduled for complete elective repair underwent palliation due to intraoperative findings of double-outlet right ventricle with possible subaortic stenosis, and repair was deferred until after cardiac catheterization. The median age at surgical repair was 8.1 months (range, 3.7 to 19.3 months), with repair at an age older than 12 months in 5 patients.

Pulmonary Valve

Palliated patients with TOF/PS had significant PV hypoplasia at baseline, with no significant improvement in Z score when reassessed immediately before complete surgical repair (Table 2 and Fig 2). However, there was statistically significant absolute growth of the PV over the time from the baseline assessment to the surgical repair ($p < .001$).

On direct operative inspection, 18 of 31 palliated patients (58%) had a bicuspid PV. The mean PV diameter in these 31 patients was 4.9 ± 1.9 mm. In our cohort, the PV was no more hypoplastic at baseline in patients with bicuspid valves (mean Z score, -2.9 ± 0.7), than in those