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Original article

Surgical strategy and outcomes for the delayed diagnosis of pulmonary atresia with intact ventricular septum

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ABSTRACT

Background: In the present study, we summarize the experiences and evaluate clinical outcomes for the delayed diagnosis of pulmonary atresia with intact ventricular septum (PAIVS) patients when undergoing an initial visit and diagnosis in our heart center.

Methods: Fifty-eight cases of delayed diagnosis of PAIVS in patients aged more than 6 months between January 2006 and June 2016 were reviewed in our hospital. The median age at initial diagnosis was 12.2 months (range, 6.1–79.6 months). Forty-five cases eventually reached definitive repair. Survival, risk factors for death, and clinical status after definitive repair were assessed.

Results: Among patients who completed definitive repair, the Fontan procedure was performed in a large proportion of older PAIVS children (42.2%, 19/45), while only a few patients received biventricular repair (22.2%, 10/45). The medium-term (10-year) survival rates of biventricular repair, 1.5-ventricular repair, and univentricular palliation were 100.0%, 93.3%, and 81.2%, respectively. At the latest follow-up, most patients had a good clinical status after definitive repairs, with a low re-operation rate.

Conclusions: A large proportion of the delayed diagnosis of PAIVS patients had to receive univentricular palliation because of limited potential for right ventricular growth. However, optimal definitive repairs could also have been achieved in these patients with a low mortality rate.

Introduction

Pulmonary atresia with intact ventricular septum (PAIVS) is a rare and complex congenital heart defect with abnormalities of the anatomical structure of the right ventricle (RV), pulmonary valve (PV), and tricuspid valve (TV) [1,2]. PAIVS remains challenging to manage because of the morphologic heterogeneity of the RV and associated abnormalities [3–6]. In current reports, the surgical treatment of PAIVS mainly focuses on neonates and younger infants, since patients without RV-dependent coronary circulation (RVDCC) have greater opportunity for subsequent biventricular repair through early RV decompression by means of surgical or catheter treatment [7–10]. Less attention has been paid to surgical treatment in delayed diagnosis PAIVS patients with older ages.

To the best of our knowledge, older PAIVS patients with a delayed diagnosis and no surgery predominantly manifest poor RV development, stable patent ductus eriosus or combined with collateral vessels, depressed heart function, and more severe cyanosis owing to the absence of communication between the RV and the pulmonary trunk. Moreover, the most significant characteristic of these patients is that the growth potential of the RV gradually decreases with increasing age [11]. Hypertrophied myocytes, diffuse fibrosis, and abnormal capillary distribution are present in the hearts of patients with PAIVS, and disordered microcirculation would continue without the palliative surgery of RV decompression [12]. In fact, the dimensions of the RV and TV relative to body size decrease as age increases in patients without reconstruction of the RV to pulmonary artery continuity [13]. The optimal surgical strategy may be different in this special population; however, to date, the clinical strategy and outcomes

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of these older PAIVS patients remain deficient in comparable studies.

In the present study, we reviewed our experience in the treatment of delayed diagnosis PAIVS patients aged more than 6 months at initial diagnosis in our heart center. We analyzed risk factors for mortality and clinical outcomes, and sought to identify appropriate approaches in terms of types of definitive repair with which to manage these older PAIVS children.

Methods

Study design and patient population

This study was approved by the ethics committee of Shanghai Children's Medical Center. Between January 2006 and June 2016, a total of 137 patients were treated for PAIVS in our heart center. Among them, 58 cases of PAIVS aged more than 6 months receiving an initial visit and diagnosis were surveyed. The median age at initial diagnosis was 12.2 months (range, 6.1–79.6 months), and the median weight at initial diagnosis was 8.2 kg (range, 4.5–17.5 kg). A diagnosis of PAIVS was based on preoperative echocardiography, partially combined with 64-slice spiral computed tomography (CT), magnetic resonance imaging (MRI), or cardiovascular angiography. The clinical characteristics of these patients are listed in Table 1. Only patients who completed definitive repairs were analyzed and evaluated in the present study.

Morphologic characteristics

Bull and associates classified the degrees of RV hypoplasia according to the tripartite, bipartite, and unipartite morphology of the RV cavity [14]. Moreover, TV *Z*-score was also proven to be closely related to RV development [15]. Based on the classification by Bull et al., the degree of RV hypoplasia in this cohort of patients was divided into three grades: mild, moderate, or severe hypoplasia according to our clinical experience and the clinical factors included, but were not limited to a combination of RV morphology, TV *Z*-score, RV cavity, tricuspid annulus diameter, or coronary artery anatomy. The TV *Z*-score was calculated as described by Hanley et al. [16]. The coronary artery anatomy was identified from the coronary angiograms. Therefore, the classification was simply described as follows: mild hypoplasia mainly with tripartite morphology and TV *Z*-score above −2, moderate hypoplasia mainly with bipartite anatomy and TV *Z*-score between −2 and −4, severe hypoplasia only with unipartite anatomy and TV *Z*-score below −4, or with RVDCC. In the present study, the 58 patients were classified into three types based on the above criteria: mild (n = 5), moderate (n = 33), or severe hypoplasia (n = 20). RVDCC was not found in any of these older children.

Table 1
The clinical characteristics of older PAIVS patients.

Demographic	*N* (%) or Mean ± SD/IQ
No. of patients	58
Gender, male/female	34/24
Weight at initial diagnosis (kg)	9.0 ± 2.8
Age at initial diagnosis (months)	18.9 ± 7.9
PDA (%)	58 (100%)
Atrial septal defect or patent foramen ovale (%)	58 (100%)
Tricuspid regurgitation (%)	45 (100%)
None or trivial	23 (40.0%)
Mild	10 (15.6%)
Moderate	13 (24.4%)
Severe	12 (20.0%)
Membranous atresia (%)	44 (75.9%)
Outflow tract muscle atresia (%)	14 (24.1%)
Tricuspid atresia (%)	1 (1.7%)
Pulmonary dysplasia (%)	2 (3.4%)
RV hypoplasia	58 (100%)
Mild	5 (8.6%)
Moderate	33 (56.9%)
Severe	20 (34.5%)
TV *Z*-score	−2.6 ± 1.5
RVDCC	0
LVEF% >50%	55 (94.8%)

PAIVS, pulmonary atresia with intact ventricular septum; PDA, patent ductus arteriosus; RV, right ventricle; TV, tricuspid valve; LVEF, left ventricular ejection fraction; RVDCC, right ventricular-dependent coronary circulation.

Surgical management

Excluding 13 cases who remained unresolved, a total of 45 PAIVS cases reached definitive repair, and were categorized into three groups as follows: biventricular repair group (2-V group), 1.5-ventricular repair group (1.5-V group), and univentricular palliation group (1-V group). These patients were treated with definitive repairs, including biventricular repair, 1.5-ventricular repair, and Fontan procedure, with or without any primary intervention. ASD closure was not recommended in these patients. In 2-V or 1.5-V group, ASD would be kept 3–4 mm if ASD was large, and small ASD does not need to be dealt with. The patients received an initial intervention based on the approaches described elsewhere, including right ventricular outflow tract reconstruction (RVOTR) with Blalock–Taussig (B–T) shunt, B–T shunt alone, or bidirectional Glenn (BDG), for example. The characteristics of the PAIVS patients managed using the different definitive repairs are shown in Table 2.

Follow-up

Different parameters, such as age at initial diagnosis, weight at initial diagnosis, TV *Z*-value, RV hypoplasia (mild, moderate, severe), definitive repair, cardiopulmonary bypass (CPB) time, intensive care unit (ICU) time, and ventilation time were analyzed as risk factors for death in PAIVS. All PAIVS survivors received echocardiography, not only for evaluation of the potential growth of the RV but also for the degree of tricuspid regurgitation and left ventricular ejection fraction (LVEF). Follow-up data were also reviewed for long-term complications and clinical status after definitive repair, including New York Heart Association (NYHA) class, arrhythmia, ventricular dysfunction, neurologic event, and need for re-operation, for example.

Statistical analysis

Statistical analysis was performed with SPSS 16.0 statistical software (SPSS Inc., Chicago, IL, USA). Mean ± standard deviation (SD) was used to describe the data. A comparison of the different groups was performed using analysis of variance (ANOVA) and least-squares difference (LSD). Actuarial survival was estimated using the Kaplan–Meier method. Risk factors for mortality were evaluated by multivariate logistic regression analysis. A p-value < 0.05 was considered to be statistically significant.

Results

Characteristics of PAIVS patients in the different groups

As shown in Table 2, 10 cases (22.2%, 10/45) with mild (50.0%, 5/10) or moderate (50.0%, 5/10) RV hypoplasia received biventricular repair in the 2-V group. Only a small proportion of older patients had the opportunity for the final biventricular repair. In the 1.5-V group, 16 cases (35.6%, 16/45) with moderate-to-severe