Echocardiographic determinants of successful balloon dilation in pulmonary atresia with intact ventricular septum

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Aims

Pulmonary atresia with intact ventricular septum (PA–IVS) is a complex congenital heart malformation with multitude therapeutic approaches. Recently, balloon valvotomy has been used as an alternative to primary surgery. This study aimed to identify echocardiographic markers of balloon dilation success in PA–IVS.

Methods and results

The echocardiograms of 26 patients diagnosed with PA–IVS who underwent primary pulmonary balloon valvotomy were reviewed. Tricuspid annulus Z-score, pulmonary annulus Z-score, right ventricular (RV) to left ventricular (LV) length ratio, RV to LV transverse diameter ratio, and tricuspid valve (TV) to mitral valve (MV) annulus diameter ratio were measured. The tricuspid Z-score, pulmonary Z-score, RV/LV length ratio, RV/LV diameter ratio, and the TV/MV ratio were significantly different in the group which had successful balloon dilation compared with that failed. Based on decision trees using the Weka classifier package, only RV/LV diameter ratio predicts a 92.3% success rate. In contrast, an RV/LV diameter ratio C 0.76 associated with RV/LV length ratio C 0.70 predicts 100% failure.

Conclusion

Successful balloon dilation in membranous type PA–IVS can be predicted by a scoring system using RV/LV diameter ratio and RV/LV length ratio.

Keywords

Pulmonary atresia • Intact ventricular septum • Percutaneous valvotomy • Echocardiography

Introduction

Pulmonary atresia with intact ventricular septum (PA–IVS) is a rare complex congenital heart malformation. It is characterized by a wide morphologic diversity in the degree of right ventricular (RV) hypoplasia, infundibular pathoanatomy, and the occurrence of right ventriculo-coronary connections. These variations influence the therapeutic approach. The goals of early palliation include the relief of cyanosis and ductal dependence by providing a reliable source of pulmonary blood flow, and the relief of RV outflow tract (RVOT) obstruction to encourage forward flow and growth of right-sided structures. How this is accomplished may vary depending on the anatomy and physiology, as well as physician and institutional bias.

PA–IVS can be difficult to manage, and surgical procedures for this anomaly have been associated with high mortality and morbidity.1 Various factors limiting the subsequent surgical outlook include size and morphology of the RV, size and function of the tricuspid valve (TV) annulus, and the presence or absence of the RV-dependent coronary circulation (RVDCC). Recently, interventional catheterization has emerged as the primary treatment for favourable forms of PA–IVS.2–6

This procedure is feasible using the stiff end of a guidewire and a laser or radiofrequency (RF) perforator, but it carries significant periprocedural risks.2,7 Recently, several authors have shown that balloon valvotomy alone for PA–IVS rarely obviates the need for an additional source of pulmonary blood flow.8,9 Further, although balloon dilation may be an alternative to primary surgery in selected candidates, it cannot be applied to all patients with PA–IVS. Currently, we believe that patients with a patent infundibulum in the setting of a non-RVDC is candidates for balloon dilation. However, the high number of procedural failures and the need for an adjunct procedure [e.g. Blalock–Taussig (BT) shunt or patent ductus arteriosus stent] argue strongly for additional studies that

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will help establish the criteria for success using balloon dilation. We therefore reviewed our experience using RF-assisted valvotomy using balloon valvuloplasty in the management of neonates and infants with PA–IVS and sought to identify the appropriate anatomic criteria for patient selection for successful balloon dilation.

Methods

With approval of our Institutional Review Board, we retrospectively reviewed the records of all patients admitted to King Faisal Specialist Hospital and Research Center with a diagnosis of PA–IVS from January 2000 to January 2007. From ultrasound recordings available in every patient, intracardiac dimensions were measured off-line using Xcelera * software (version 1.2 L4, Philips) by a single investigator (A.D.) blinded to treatment outcome. Morphologic study parameters in the apical four-chamber view at end-diastole included RV and LV lengths taken from the mid-portion of the atrioventricular annuli to the apex of the RV and LV, RV and LV maximal diameters measured just below the TV and mitral valve (MV) leaflets tips with the tips open, and TV, pulmonary valve (PV), and MV annular diameters in diastole (Figure 1). We classified RV morphology according to the mono-, bi-, and tripartite nature of the ventricle cavity as reported by Bull et al.10 and the type of PA–IVS (as muscular or membranous). We considered RVDCC to be present when fistulous communications were associated with either absent aorto-coronary connections, coronary artery interruption, or unequivocal stenosis of one or more of the major epicardial coronary arteries as determined by coronary angiography. Each patient had root or individual coronary angiography.

Patients

During the study period, 46 patients, 26 of whom were male, with a median age at diagnosis of 6 days (range 1–49 days) with PA–IVS were admitted to our institution, and 26 underwent RF-assisted pulmonary valvotomy with balloon pulmonary valvuloplasty. The other 20 patients underwent first-line surgery. The treatment plan was based on discussion among surgeons, cardiologists, and intensivists taking into account RV and TV size, morphology and status of the coronary circulation, and morphologic type of PA–IVS. Patients were not randomly assigned to treatment, and no defined institutional protocol was followed for stratification. All patients had prostaglandin E1 (PGE1) infusion prior to and during surgical or catheter intervention.

Patients with a diminutive, severely hypoplastic RV and TV annulus (TV annulus Z-score less than –3) as well as absence of an RV infundibulum were excluded and were assigned for eventual single-ventricle repair.

Transcatheter primary procedure

Twenty-six patients underwent cardiac catheterization under general anesthesia and were subjected to RF valvotomy and balloon dilation. The femoral artery and vein were cannulated percutaneously using 4 and 5 F sheaths, respectively. Heparin (100 UI/kg) was given intra-arterially once intravascular access was achieved. RVDCC was first excluded in all patients by coronary angiography. RF valvotomy was then accomplished using an RF generator (Model 1025A-115, Baylis Medical Company, Montreal, Canada). Balloon dilation was performed sequentially using 2 and 4 mm coronary balloons and finally 8–10 mm balloon valvuloplasty. Successful balloon dilation was defined as the ability to puncture the valve plate with a wire, inflate the balloon to the size of the valve annulus, and subsequently wean from prostaglandin with sustained room air oxygen saturations ≥70%.

Using these criteria, 13 of 26 patients had successful balloon dilation (Group I). The mean PGE1 duration after the procedure was 8.9 ± 5.3 days (range 1–19 days) and the RVOT gradient was 24.6 ± 7.8 mmHg. Thirteen patients had a technically unsuccessful balloon dilation, due either to failure to wean from prostaglandin or to persistent cyanosis (<70%) off PGE1 (Group II); 12 patients in this group underwent placement of a systemic-to-pulmonary (BT) shunt. One patient underwent pulmonary valvectomy and placement of an RVOT patch without a BT shunt. No death was recorded in the two groups. We noted two cases of acute renal failure, two cases of low cardiac output syndrome, and one case of septicemia in Group I and one case of acute renal failure, one case of low cardiac output syndrome, two cases of endocarditis, and one case of pleural effusion in Group II. All complications were resolved without sequelae.

Statistics

Data are expressed as mean ± SD. Baseline values between groups were compared using Mann–Whitney U-test and Fisher’s exact test. We used classification of clinical data using decision trees.3,11 The Decision Tree is one of the most popular classification algorithms in the current use in Data Mining and Machine Learning. In the process of the construction of decision trees, the variables are sorted out according to the quantity of information each contains relative to the decision or class. This quantity of information is computed using the Shannon entropy measurement. The decision tree is built from its root to its leaves. The training set is successively split by means of questions on the value of a chosen variable. The obtained decision tree is interpreted as a set of rules (i.e. if.then.) generalizing the training set. For the experiment, we used the J48 decision tree in the Weka (www.cs.waikato.ac.nz/ml/weka/) data mining package version 3.

Figure 1 Anatomic parameters that were measured in the cardiac four-chamber view.
A training set of 26 subjects is constituted and described by 33 variables; class is coded by a binary variable indicating success or failure of balloon dilation.

### Results

#### Comparison of findings at diagnosis

Data derived from baseline demographic and echocardiographic characteristics are listed in Tables 1 and 2. There were no significant differences in age at diagnosis, age at first procedure, PG duration, or length of intensive care unit and hospital stay. In both groups, there was no RVDCC. TV/MV ratio, RV/LV length ratio, RV/LV diameter ratio, TV annulus, TV Z-score, PV annulus, PV Z-score, and RV length Z-score were significantly different between Groups I and II (Table 2).

The decision tree algorithm resulted in the obtained following rules (Figure 2):

Rule 1: If RV/LV diameter ratio is >0.76, the probability of successful balloon angioplasty is 92.3%.

Rule 2: If RV/LV diameter ratio is ≤0.76 and RV/LV length ratio ≤0.70, then the probability of an unsuccessful balloon angioplasty is 100%.

Rule 3: If RV/LV diameter ratio is ≤0.76 and RV/LV length ratio >0.70, then the probability of successful balloon angioplasty is 75%.

#### Follow-up

The median follow-up (FU) was 9 months (range 0–60 months). In Group I, six patients had a second balloon valvuloplasty associated in two patients with patent foramen ovale/atrial septal defect device closure. The mean age at the second procedure was 11.8 ± 8.7 months.

In Group II, three patients had biventricular repair, one patient had one and half ventricular repair, three patients had Fontan procedure, and one patient died during FU. Five patients are waiting for surgery.

### Discussion

PA–IVS represents a congenital heart malformation with a broad spectrum of morphologic heterogeneity. At one end of the spectrum are those patients with mild-to-moderate hypoplasia of the RV who generally have a patent infundibulum. At the other end of the spectrum are those with severe hypoplasia of the RV who often have major RVDCC. For patients in the first group, the treatment goal is to eventually establish a complete biventricular circulation with RV decompression as the initial procedure. Patients
with a severely hypoplastic, diminutive RV are generally assigned to a univentricular repair. Recently, the use of a laser wire and RF-assisted valvotomy and balloon dilation has made possible primary treatment of this group of patients in the catheterization laboratory. However, the appropriate anatomic criteria for patient selection for catheter intervention have not been clearly established. In a recent editorial, Cheatham suggested that as long as there is a tripartite RV and a well-formed infundibulum, a TV annulus $\geq 11$ mm, and membranous atretic PV $\geq 7$ mm, transcatheter therapy should be the preferred therapy. However, no supportive data for these recommendations were given. World-wide there is no consensus as to the best approach to open the RVOT in subjects who have the membranous type of PA–IVS with a tri- or bipartite RV in the setting of a non-RVDCC, and there are no comparative studies regarding the best initial approach. Nevertheless, a wide range of reported algorithms exist for management. To the best of our knowledge, our approach. Nevertheless, a wide range of reported algorithms exist for management. We thank Mr Hamlich Mohammed for his statistic help.

Conclusion

Transcatheter valvotomy and balloon dilation offer a promising alternative as the primary therapy in selected patients with PA–IVS. We believe that simple ECHO markers that include RV/LV diameter ratio and RV/LV length ratio can help in determining a priori which patients will benefit from transcatheter pulmonary valvotomy in patients with PA–IVS.

Acknowledgement

We thank Mr Hamlich Mohammed for his statistic help.

Conflict of interest: none declared.

References

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Study limitations

The most important limitations were the retrospective study design and the relatively small sample size. The small sample size is due in part to the rarity of this disease and by the relatively infrequent use of valve plate perforation and balloon dilation as the primary therapeutic approach. Our sample size is no different from that reported in most published series.