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ORIGINAL ARTICLE

CONGENITAL

Pulmonary artery banding as 'open end' palliation of systemic right ventricles: an interim analysis

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Abstract

OBJECTIVES: A morphological right ventricle (RV) is not ideally suited for the long-term maintenance of the systemic circulation. The aim of this analysis was to evaluate the intermediate results and outcome of pulmonary artery banding (PAB) in an open-ended strategy.

METHODS: This is a retrospective review of patients with systemic RVs who had undergone PAB in our institution from April 1985 to January 2011. PAB was placed in 5 patients late after the Senning operation and in 15 patients with corrected transposition; of whom, 6 had a large ventricular septal defect.

RESULTS: PAB was performed at a median age of 4.3 years (range: 0.9–14.9), median follow-up of 86 months (range: 0.5–379). All 20 patients are alive and are being followed up. Tricuspid regurgitation (TR), RV function and dilation showed no deterioration after banding ($P = 0.9$). Ninety per cent (18/20) have adequate ongoing palliation with PAB. One patient underwent a double-switch operation and one received an additional bidirectional Glenn shunt. A dilatable band was redilated with improvement in percutaneous saturation and in another the procedure was abandoned due to development of transient atrioventricular block. Functional class remained either unchanged or improved.

CONCLUSIONS: PAB was performed with no mortality and low morbidity. PAB in these heterogeneous patients provides true 'open ended palliation' by allowing left ventricular training in those going for anatomical repair, stabilizing or improving RV function and TR in others, thereby delaying surgery. It can also be left in place as long-term palliation. The addition of a dilatable band allows manipulation of pulmonary flows, but longer follow-up is required to provide data on best management strategies for these complex patients.

Keywords: Congenital cardiology • Pulmonary banding • Dilatable band • Double-switch • Systemic right ventricle

INTRODUCTION

A systemic morphological right ventricle (RV) is not ideally suited for the long-term maintenance of the systemic circulation. It may fail in time under these conditions because of its anatomical and physiological substrate [1]. RV dysfunction is unpredictable and has a wide range of clinical presentations varying from overt cardiac failure in young patients to asymptomatic in the elderly. Natural evolution and the broad range of outcomes are also poorly understood.

Systemic RV's in patients with congenitally corrected transposition of the great arteries (c-TGA) and in patients after an atrial switch operation (Mustard or Senning) for d-transposition—whose physiology are comparable with those with corrected transposition—show signs of poor adaptation as patients grow older. Severe RV dysfunction will occur by the third decade in ~10% of patients following an atrial switch operation for transposition of the great arteries. This number is expected to increase with this cohort currently entering late adulthood [2, 3].

These systemic RV's may exhibit varying degrees of dysfunction, dilation and tricuspid regurgitation (TR) [1]. Current surgical strategies to deal with a failing systemic RV consist of anatomical repair (double-switch) and cardiac transplant [4–9]. Anatomical repair is considered as the 'best' option of palliation provided that the left ventricle (LV) is trained, which is the case in infants and neonates or after PAB in childhood [7, 10].

Pulmonary artery banding (PAB) has been used as an interim measure either to prevent pulmonary hypertension, stabilize TR or to train the LV [11, 12]. However, the use of a fixed band during childhood may force the physician to proceed to early anatomical repair if the band becomes too tight, even in the presence of good RV function. In the past couple of years, dilatable bands have become available [13–15]. The advantage is that if RV dysfunction improves after banding, surgery can be delayed or, alternatively, if the band becomes too tight, it can be partially released by simple angioplasty.

We embarked on a strategy of PAB in some patients with systemic anatomical RV's. Patients were banded because of RV

dysfunction, hoping that RV function would improve sufficiently to stabilize or improve the patients and to have the LV trained for double-switch operation if required. The aim of this study was to evaluate the intermediate results of the effectiveness of this open-ended strategy.

PATIENTS AND METHODS

This was an interim, retrospective review of patients with systemic morphological RV's who had undergone PAB in our institution from April 1985 to January 2011. PAB was performed in 20 patients who had systemic RV's. Patients were divided into three groups according to main diagnosis and reason for PAB (details in Fig. 1). Indications for banding varied widely. In the Senning group ($n = 5$), it was used concomitant to elective surgery (residual atrial septal defect closure: $n = 2$, epicardial pacemaker placement: $n = 1$) or to prepare the LV for double-switch ($n = 2$). In six infants with c-TGA and large ventricular septal defects (VSDs), restrictive bands were placed to control pulmonary blood flow. In asymptomatic patients with c-TGA ($n = 9$), bands were initially electively placed at the time of implantation of a sequential epicardial pacemaker. Indications were subsequently extended for prognostic reasons to include even patients with slight progression of TR and/or RV dysfunction.

Patient demographics and clinical data were obtained from our local database. Ventricular function and size were assessed by means of echocardiography. A single cardiologist reviewed all previous echocardiograms and performed independent measurements. TR and RV dilation were scored as follows: none, 0; trivial, 1; mild, 2; moderate, 3 and severe, 4. Quantitative measurements included standard M-mode LV end-diastolic and systolic measurements, two-dimensional RV end-diastolic dimensions on a four-chamber view and Doppler peak instantaneous gradients over the banding. Catheterization and radiographic information were also incorporated in the assessment if available.

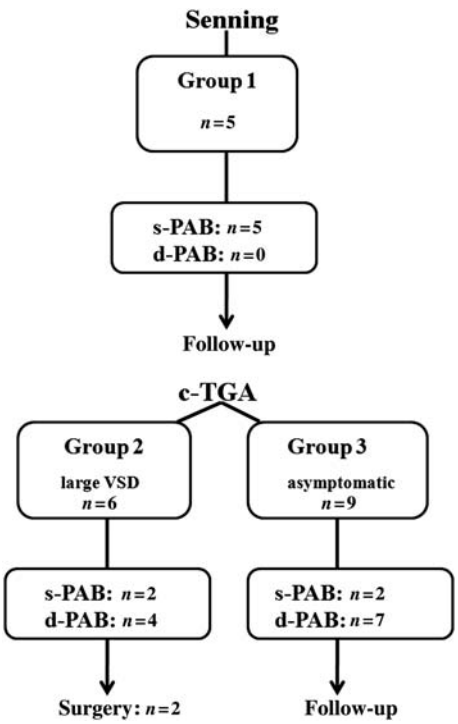

Figure 1: Classification and management of study groups. c-TGA, corrected transposition of great arteries; VSD, ventricular septal defect; s-PAB, standard pulmonary artery banding; d-PAB, dilatable pulmonary artery banding.

Early in the series, PAB was performed using standard surgical technique in nine patients. In 11 patients, a dilatable band was placed after late 2004. The dilatable band was created according to a technique we have previously described: it consists of a standard band closed with vascular clips placed at short intervals [14]. Except for those with a large VSD, the surgeons were requested to place a mild band to accommodate for growth, with the aim to result in subsystemic LV pressures when fully grown. Overall, mild banding was achieved by creating a discernible indentation of the pulmonary artery, aiming for a gradient of ~20–40 mmHg without LV dysfunction on transoesophageal echocardiography.

Data were analysed using standard statistical software (SPSS for windows, SPSS Inc., IBM company, Chicago, IL, USA, version 18). A paired Student's *t*-test or analysis of variance was used to compare normally distributed data. Skewed data were analysed non-parametrically using the Kruskal–Wallis test. Continuous data were expressed as medians with minimum and maximum values where appropriate.

Ethical clearance was obtained from the local ethics committee. Informed consent was obtained in all from either the parents, legal guardians and/or from the patients where appropriate.

RESULTS

The study group consisted of 12 males and 8 females. PAB was performed at a median age of 4.3 years (range: 0.1–14.9). Patient characteristics can be viewed in Table 1. All patients are alive and all are being followed up with a current median follow-up

Table 1: Patients characteristics

	Senning	c-TGA with VSD	c-TGA
Number (*n*)	5	6	9
Age at PAB (years)			
Median	7.7	0.2	4.3
Range	5.4–11.9	0.1–0.6	0.9–14.9
Follow-up after PAB (months)			
Median	153.9	71.6	12.5
Range	136.1–162.5	9.6–379.4	0.5–84.5
PIG LVOT (mmHg)			
Immediately after banding			
Median	15	70	38
Range	10–35	40–95	22–52
Last follow-up			
Median	30	79	40
Range	16–67	70–85	30–70

Patient characteristics and left ventricular outflow tract (LVOT) gradient follow-up. PAB, pulmonary artery banding; PIG, Doppler peak instantaneous gradient.

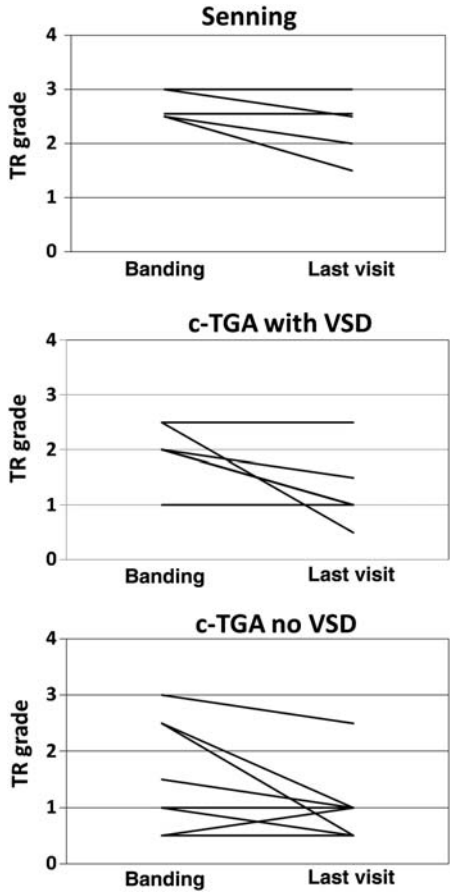

Figure 2: Progression of TR. TR grade immediately after banding and at last visit.

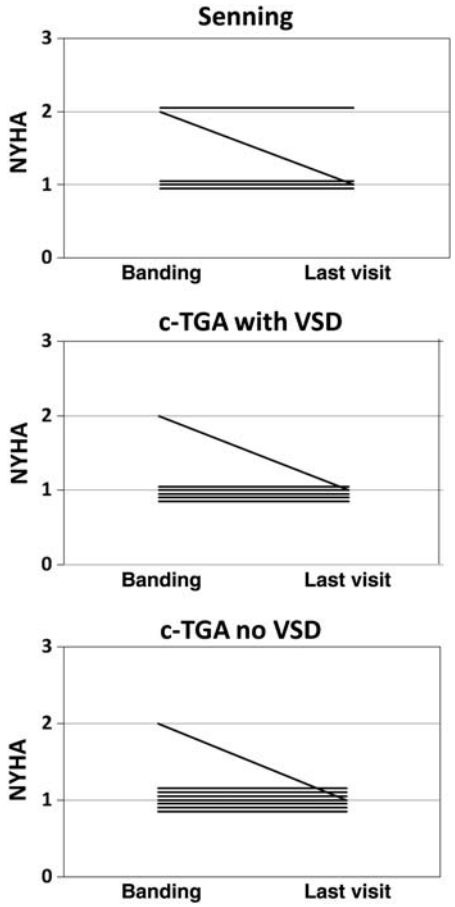

Figure 3: Progression of NYHA functional class. Functional class immediately after banding and at last visit.

of 86 months (range: 0.5–379). Two patients in the Senning group and six children in the corrected transposition group had pacemakers implanted.

The gradient over the LV outflow tract showed a mild but non-significant increase over time ($P = 0.90$; 0.11 and 0.08 for the groups, respectively) (Table 1). RV function and dilation showed no significant deterioration during the follow-up period. RV function improved in one patient in Group 1 and one in Group 2. Improvement in NYHA functional class and TR can be viewed in Figs 2 and 3 and the difference of change in Table 2. Nonetheless, these differences were not statistically significant. No changes were observed in LV end-systolic diameters. None of the patients showed a deterioration of functional status after PAB. In the Senning group, three patients were in NYHA functional class I and two in II before PAB and afterwards four were in functional class I. In the group with VSD's, five were in class I and one in class II; all were in functional class I on follow-up. Eight of the asymptomatic group were in functional class I and one in class two, all were in class I during follow-up (Fig. 3).

Table 2: Difference of change in functional class and TR after PAB

	Variable	Median	Difference of change		
			Mean	Minimum	Maximum
Senning	NYHA	0	−0.2	−1	0
	TR	−0.5	−0.4	−1	0
c-TGA, large VSD	NYHA	0	−0.17	−1	0
	TR	−0.7	−0.7	−2	0
c-TGA, no VSD	NYHA	0	−0.1	−1	0
	TR	−0.5	−0.5	−2	0.5

NYHA, New York Heart Association functional class; TR, tricuspid regurgitation; VSD, ventricular septal defect.

Outcome and complications

One patient had transient LV dysfunction due to LV pressure overload and one had diaphragmatic paresis after placement of the PAB, both of which normalized spontaneously within 6 and 16 months, respectively.

One patient with a dilatable band received angioplasty due to progressive cyanosis 14 months after PAB placement. Percutaneous saturations improved from 85 to 93%. In another, pulmonary angioplasty of a dilatable band was aborted due to development of temporary atrioventricular block; with adapted technique, this might be avoidable, but the patient progressed to an additional Glenn shunt. No other patients have yet required balloon angioplasty of the band (Fig. 4).

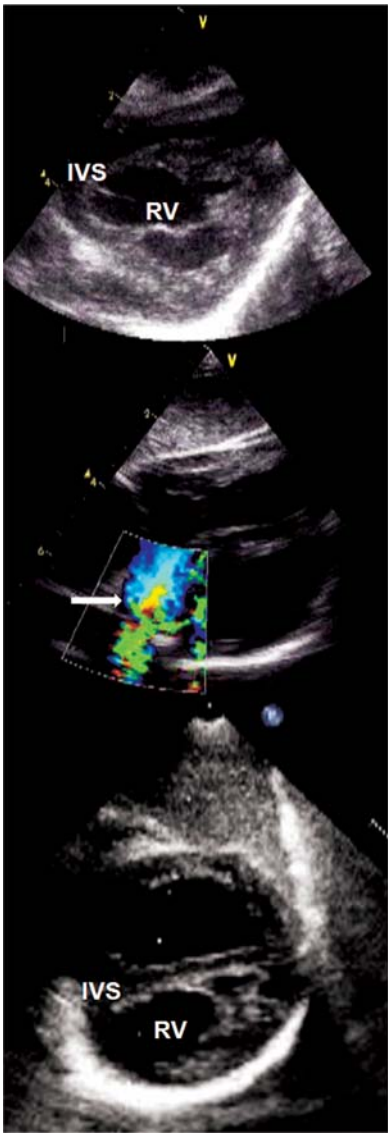

Figure 4: Echocardiographic follow-up subcostal views of a patient with c-TGA. Top, pre-banding, note dilation of the RV and position of the interventricular septum; middle, turbulence over banding (arrow); bottom, the nice shape of the interventricular septum; expansion of LV.

PAB provided continuous adequate palliation in 90% (18/20) of the patients. Two patients in Group 2 had additional surgery. One patient with a fixed band proceeded to anatomical repair and in the child with temporary atrioventricular block and a borderline LV, a bidirectional Glenn with pulsatile pulmonary flow was carried out.

DISCUSSION

The clinical progression of patients with c-TGA physiology is extremely variable and unpredictable. Natural history studies have reported survival of 70 and 64% at 5 and 10 years after diagnosis, respectively, with RV failure and TR increasingly leading to cardiac failure [16]. Many clinicians are disgruntled by current management strategies and as a result, this topic is frequently encountered at conferences and meetings.

Once ventricular dysfunction and cardiac failure occurs, typically after the second decade, the clinician is left with limited options. Medical treatment has not yielded spectacular results. Current surgical strategies are not ideal. The LV needs to be trained and if banding is performed after puberty, several banding procedures are required with significant mortality and morbidity, while only some proceed to double-switch [16, 17]. The strategy of double-switch, although heralded as an ideal solution, does not return everything to 'normal'. Reported early mortality rates vary between 0 and 9% with freedom from re-operation of 93 and 77% at 5 and 10 years, respectively [7, 18]. Also, follow-up in surgical series is now only early to mid-term, and long-term morbidity and survival have yet to be determined. Postoperative problems include superior and inferior vena cava baffle obstruction (3%), atrial arrhythmias and neoaortic root dilation and regurgitation [19]. When confronted with acute heart failure, a ventricular assist device may be considered, but remains late and bail-out palliative treatment [20]. Cardiac transplant is also not an ideal solution with an annual attrition rate of ~2.5% [21].

Clinicians are frequently impressed that patients with c-TGA and pulmonary outflow obstruction fare better than those without. It has unequivocally been established that PAB has beneficial effects in patients with morphological RV's acting as the systemic ventricle. PAB has been demonstrated to effectively train the LV, improve RV function, regress or stabilize tricuspid valve regurgitation, preventor delay cardiac transplantation and may even be used as palliation [22–24]. In this analysis, PAB served as interim palliation in 90% of our group, indicating the effectiveness of PAB as a palliative procedure. Theoretically, PAB increases the after load of the morphological LV and effects a shift of the interventricular septum back towards the midline position, causing the RV to become less spherical. This LV interference moves the attachment of the septal leaflet of the tricuspid valve and improves tricuspid valve co-aptation, thereby reducing regurgitation [22, 25]. As a result, RV preload is reduced with improvement of RV dilation and systolic function [3].

Many cardiothoracic surgeons are reluctant to use PAB as a form of treatment because they find it difficult to decide on what is 'tight' enough while others regard it as inferior compared with a double-switch operation. However, in our limited experience, PAB has proven to effect a true 'open ended' palliation: it can be used as long-term palliative treatment or act as a bridge to reparative surgery or transplantation. Our results have shown that PAB has no early mortality, low morbidity and stabilizes RV dysfunction as well as TR. Also, patients remain in good functional NYHA classes. This is in agreement with the findings of other studies [18, 23].

Fixed bands may become too tight during growth and one may be forced to either adjust the band and/or proceed to double-switch. The concept of dilatable bands later in our experience added a new dimension. This method of adjustable banding allows further manipulations based on distal movement of the clips with sequential angioplasty [14]. Pulmonary flow can be enhanced and this was attempted in two patients. In one, significant improvement in percutaneous saturation was obtained, but the other developed a transient heart block during the procedure and it was thus aborted. This problem might be avoided with adapted technique.

In the subgroup of patients with corrected TGA and large VSD's, we were pleased with the results of PAB. None of the patients developed pulmonary hypertension during follow-up

and one progressed to anatomical repair without the need for a pacemaker. The other patient was a child who developed a heart block during attempted dilation of PAB; since the LV volume was also borderline, a bidirectional Glenn was performed as 'final' palliation. Delaying surgery in this group is especially helpful since surgical options are limited and associated with high morbidity: classical closure of VSD or double-switch yields a significant risk of atrioventricular block and total pacemaker dependency; lack of escape rhythm may lead to death in the case of lead fracture or exit block and epicardial leads may cause coronary strangulation in young, growing children. In this group, especially in the presence of a borderline LV, an elegant alternative to consider is a one-and-a-half repair, as performed in the abovementioned child.

Optimal timing and degree of PAB remain difficult questions to answer. In an untrained LV, it is difficult to predict what the response will be to increase in afterload; also, age and the underlying cardiac defects make it difficult to decide what is 'tight' enough [17]. It is evident that there is an age limit: if banding is delayed until after puberty, it will be too late resulting in inadequate training of the LV. This can be explained on the basis of reduced coronary flow reserves where the rapid LV hypertrophy following banding may induce subendocardial ischaemia. Poirier and Mee [24] showed myocardial oedema on magnetic resonance imaging shortly after banding which is believed to be a precursor of fibrosis.

Metton *et al*. [13], on the other hand, proposed early aggressive infantile banding. Research has shown that in infants, the increase in LV mass is due to hyperplasia rather than hypertrophy and one can only speculate that less long-term problems with LV dysfunction may thus be expected. However, a PAB applied in early infancy may become too tight too soon. We prefer mild banding in small children, allowing them to 'grow' into their bands. c-TGA with VSD should be banded at an early age (<3 months), while in asymptomatic patients, PAB can be delayed into early childhood to be performed electively or when an opportunity arises (e.g. pacemaker implantation). Most Senning patients are now beyond puberty and should be banded as tight as possible, which in our experience is usually at best mild to moderate.

Unanswered concerns remain. If PAB is prophylactically performed, how will functional capacity and exercise tolerance be influenced in the banded state? In patients who would not have developed RV dysfunction, can PAB have a deleterious effect? When is the LV adequately trained and what effects can PAB have on the neo-aortic valve? Although this study does not provide the answers, our clinical impression is that moderate PAB is beneficial. Also, PAB does not disqualify a patient from anatomical repair. As a matter of fact, corrective surgery should be performed before RV failure sets in; good RV performance is essential for acceptable long-term outcomes.

Limitations

Although this is an ongoing study, this interim analysis is retrospective in nature. Numbers are small, making statistical analysis of small changes difficult. Evaluation has mostly consisted of serial echocardiographic findings. In order to reduce observer bias and variability, all echocardiograms were independently re-examined and scored by a single cardiologist. Magnetic resonance would have produced more objective measurements and is included in the future follow-up of our patients. It is evident that long-term follow-up and data are imperative.

CONCLUSION

PAB is safe and effective in patients with systemic morphological RV's. PAB in these heterogeneous patients provide true 'open ended palliation' by allowing LV training in those going for anatomical repair while improving or stabilizing RV function and TR in others. It may also be left in place as long-term palliative treatment in small children. The addition of a dilatable band allows manipulation of pulmonary flows, but longer follow-up is required to provide data on best management strategies for these complex patients.

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