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ORIGINAL ARTICLE

Tetralogy of Fallot repair: optimal *z*-score use for transannular patch insertion

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CONGENITAL

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Abstract

OBJECTIVES: Transannular patching is used to relieve significant pulmonary annular stenosis during tetralogy of Fallot repair. Although recent literature has focused on the deleterious effects of pulmonary regurgitation, inadequate relief of stenosis may increase post-operative mortality and the re-intervention rate. Patching criteria based on pulmonary annular *z*-scores are ambiguous because of the use of varied *z*-score data sets. This study aimed to generate data that could be used to optimize *z*-score use for patch insertion.

METHODS: A retrospective review was made of medical records of patients who had a valve-sparing repair of tetralogy of Fallot between 1 January 2000 and 31 December 2010. In a selected group in which the residual gradient was confined to the pulmonary valve, the post-repair peak pressure gradient was determined by trans-thoracic echocardiography and was correlated with the intra-operative pulmonary valve annulus (PVA) diameter *z*-score. Regression analysis was used to examine this relationship.

RESULTS: Data from 46 patients with valve-sparing repair were reviewed; the median age and median weight were 6.5 months and 6.6 kg, respectively; the 30-day mortality was 2.2%. Analysis of these data implies that 25% of the time, all patients with a PVA *z*-score of −1.3 would have a PVA gradient ≥30 mmHg.

CONCLUSIONS: Criteria that recommend a transannular patch insertion only when the PVA diameter *z*-score is significantly smaller than −1.3 (e.g. −3) may result in a significant number of patients with an unacceptable post-repair PVA gradient.

Keywords: Paediatric • Congenital heart disease • Pulmonary valve • Practice guidelines

INTRODUCTION

The fundamental components of surgical correction of tetralogy of Fallot (TOF) are ventricular septal defect closure and relief of significant right ventricular outflow tract obstruction (RVOTO). Relief of significant pulmonary annular obstruction is achieved by the interruption of the pulmonary valve annulus (PVA) and the insertion of a transannular patch (TAP). Although right ventricular dilation and dysfunction are important late consequences of free pulmonary regurgitation, there is evidence that failure to insert a TAP when one is indicated increases surgical mortality [1].

Although numerous criteria exist for TAP insertion, there is no agreement on which is the best. Early work resulted in the development of a table of 'minimal acceptable' pulmonary valve ring diameters to aid surgical decision-making [2, 3]. Alternative decision-making aids are based on PVA *z*-scores [4–7]. None of these criteria objectively indicate the likelihood of having an unacceptable postoperative pressure gradient across the PVA.

A previous report from our centre discussed the impact of the variation in normative scoring for cardiac structures in paediatric cardiac surgical practice and recommended an optimal normative dimension data set (that of Pettersen *et al.*) [8]. We are concerned that the inappropriate use of *z*-scores in surgical decision making may adversely affect operative outcomes [9]. Our study was undertaken to examine the relationship between the PVA diameter *z*-score and the postoperative trans-pulmonary gradient using a local contemporary clinical data set. These data could then be used as a pilot group to help refine surgical decision making with respect to TAP insertion.

PATIENTS AND METHODS

To analyse the relationship between the gradient across the PVA and the PVA diameter *z*-score, a retrospective review was made of medical records of patients who underwent repair of TOF (at the Green Lane/Starship Paediatric and Congenital Cardiac Surgery service, Auckland, New Zealand) between 1 January 2000 and 31 December 2010. The post-repair velocity across the PVA was determined by trans-thoracic echocardiography (after removal of pacing wires; generally on postoperative day 4). This velocity was converted to a pressure gradient across the PVA using the standard modification of the Bernoulli equation $P = 4v^2$ (where P is the pressure gradient across the PVA and v is the velocity of blood flow across the PVA measured at

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echocardiography). The gradient was correlated with the PVA diameter *z*-score determined intraoperatively using Hegar's dilators (avoiding over distension of the PVA) and by utilizing *z*-scores derived from the work by Pettersen *et al*. [8]. Fused commissures were incised and leaflets were partly mobilized off the pulmonary artery wall as required. The PVA was sized after such manoeuvres. To ensure that our data expressed the actual relationship between the PVA diameter *z*-score and the PVA gradient, we attempted to define a subgroup of patients within the tetralogy group for whom the echocardiographically determined peak velocity across the right ventricular outflow tract would define the pulmonary valvar stenosis as purely as possible. Only those patients with an interpretable Doppler signal from appropriate interrogation of the right ventricular outflow tract were included for whom pulse wave and continuous wave modalities were appropriately employed. Full inclusion and exclusion criteria are summarized in Table 1. In particular, patients with evidence of residual subvalvar and supravalvar stenosis were excluded. Patients who would require a conduit for repair were also excluded. During the period of this study, we used the TAP insertion criteria proposed by the table developed by Pacifico *et al*. [3]. All echocardiographic measurements were made using the Phillips IE 33 or Sonos 5500. Variables are expressed as the median (with ranges). All analyses were performed using SAS 9.1 software. Ethics approval for the study was given by the Northern X Regional Ethics Committee, and institutional approval was provided by the Auckland District Health Board Research Review Committee.

Table 1: Inclusion and exclusion criteria

Inclusion criteria	Exclusion criteria
1. Less than 19 years old at surgery 2. Had 'valve-sparing' repair	1. Greater than mild TR 2. Greater than moderate PR 3. Greater than mild RV dysfunction 4. SP-RVOT velocity >1.2 m/s 5. Suprapulmonary (MPA) stenosis 6. Branch pulmonary artery stenosis 7. Non-restrictive residual VSD 8. TET-PA 9. TET-APV 10. Chronic lung disease

TR: tricuspid valve regurgitation; PR: pulmonary valve regurgitation; RV: right ventricle; SP-RVOT: subpulmonary right ventricular outflow tract; MPA: main pulmonary artery; VSD: ventricular septal defect; TET-PA: tetralogy of Fallot with pulmonary atresia; TET-APV: tetralogy of Fallot with absent pulmonary valve.

Table 2: Patient demographics ($n = 46$)

Variable	Median	Range
Age (months)	6.50	1.25–93.00
Weight (kg)	6.65	3.79–17.40
BSA (m^2)	0.36	0.22–0.74
PVA (mm)	10.00	7.00–14.00
PVA *z*-score	−0.53	−1.63 to 1.17
Bypass time (min)	115.00	79.00–228.00
AXCT (min)	90.50	54.00–175.00
ICU stay (days)	1	1–7
Hospital stay (days)	7	3–20

BSA: body surface area; PVA: pulmonary valve annulus; AXCT: aortic cross clamp time; ICU: intensive care unit.

RESULTS

During the study period, 243 patients had repair of TOF; 106 (43.6%) had a valve-sparing repair (VSR). Of the VSR group, 46 (43.4%) met the criteria. Demographic details of these patients are summarized in Table 2. Median age of patients at repair was 6.5 months and median weight was 6.6 kg; the 30-day mortality was 2.2%. Twenty, 15 and 11 patients had no, trivial and mild postoperative pulmonary valve regurgitation, respectively; no patient had moderate regurgitation. Eleven patients (23.9%) had a residual ventricular septal defect (all were considered small); of these, 5 (10.9%) had pulmonary valve regurgitation as well.

A scatter plot of the PVA *z*-score against the post-repair PVA gradient was plotted. The data were subjected to linearity testing; there was little evidence against linearity. We subsequently chose a linear fit; the 'best-fit' line with the 50% predictive interval is shown in Fig. 1 ($r^2 = 0.201$; $P = 0.002$).

In our current practice, the typical patient undergoes TOF repair at 6 months of age and has a body surface area (BSA) of ~0.35 m^2. Table 3 was designed to compare the potential clinical implications (in terms of blood flow) of various TAP insertion criteria. Using the Hagen–Poisseuille equation that describes the flow of fluids in tubes, the table illustrates relative flow across the PVA when various TAP insertion criteria are employed for a patient with a BSA of 0.35 m^2. Flow across the PVA using the Pacifico table criterion is used as the base line for comparison (flow of 1.0).

DISCUSSION

In regard to RVOTO, the goal of TOF repair is to provide an adequate RVOT while minimizing surgical mortality and re-intervention for RVOTO. There is variation in practice with regard to the acceptable post-repair PVA gradient; a post-repair PVA gradient of 50 mmHg would be considered by most clinicians as unacceptable, while some clinicians feel that a gradient of 40 mmHg warrants interruption of the PVA. PVA gradients <30 mmHg are very unlikely to lead to progressive RVOTO [10]. The rationale behind our choice of 30 mmHg as a potential boundary for acceptability is 2-fold: firstly, at a gradient of greater than 30 mmHg there is a significant chance of progression of RVOTO [10]. Secondly, a gradient of 30 mmHg, in this infant population, would imply that the right ventricular systolic pressure to left ventricular systolic pressure ratio (RVSP/LVSP) would be greater than 0.5; this has been associated with increased surgical mortality [1, 11, 12]. Both these situations are unacceptable in terms of surgical mortality and the need for re-intervention for progressive RVOTO.

The current study provides a regression line that describes the relationship between the gradient across the PVA and the PVA diameter *z*-score in our cohort. By excluding patients with subvalvar stenosis or supra-valvar stenosis and trying to optimize the

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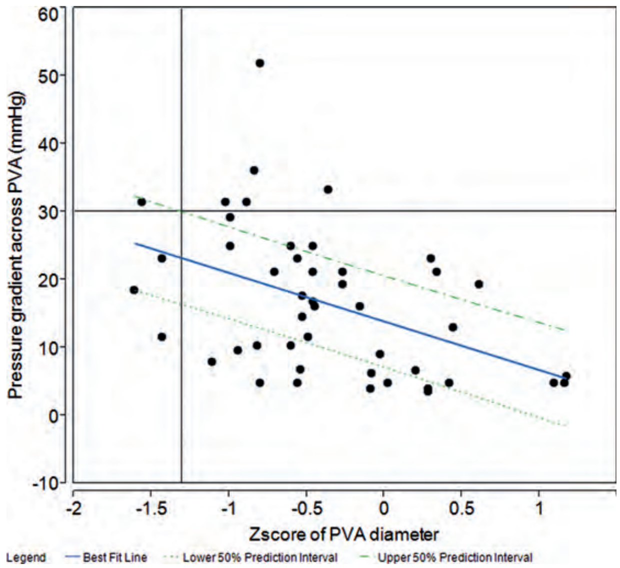

Figure 1: PVA pressure gradient vs PVA diameter *z*-score. PVA: pulmonary valve annulus.

Table 3: Comparison of criteria, PVA diameter and flow

TAP criteria	Minimal acceptable PVA diameter (mm) for BSA of 0.35	Relative flow
Pacifico table [3]	9.0	1.00
Rowlatt *et al.* [20] *z*-score of −3 [4]	6.5	0.27
Daubeney *et al.* [13] *z*-score of −3 [21]	7.8	0.56
Rowlatt *et al.* *z*-score of 0 [5]	10.0	1.52
Pettersen *et al.* [8] *z*-score of 0	10.2	1.65
Daubeney *et al.* [13] *z*-score of 0	11.1	2.31

BSA: body surface area (m^2); TAP: transannular patch; PVA: pulmonary valve annulus.

normalization or z scoring, we have endeavoured to determine a true relationship between the PVA gradient and the PVA diameter *z*-score. Figure 1 shows that 75% of the time, all patients with a PVA *z*-score of −1.3 would have a PVA gradient ≤30 mmHg. Although pulmonary valve annuli with very small *z*-scores are not examined by this model, the figure shows that, as one would expect, as the size of the pulmonary valve decreases, the likelihood of having an unacceptable gradient across the PVA annulus increases. Data from these patients imply that when the PVA *z*-score is −1.3, there will be a gradient of 30 mmHg or greater 25% of the time. Hence criteria that recommend TAP insertion only when the PVA diameter *z*-score is significantly smaller than −1 (for example, −2 or −3), may result in a substantial number of patients (>25%) with a significant post-repair PVA gradient.

The R^2 of 0.201 implies that ∼20% of the variability in the PVA gradient is explained by the PVA size. As the gradient across the PVA depends on the magnitude of the flow across the annulus, factors that affect flow across the PVA could also contribute to the variability. In this regard, varying degrees of intraoperative myocardial damage may affect the right ventricular contractility and subsequent flow across the PVA. Similarly, varying degrees of right ventricular hypertrophy and the resultant diastolic dysfunction might alter the right ventricular preload and subsequent flow across the PVA.

From Table 3, it can be seen that criteria that recommend TAP insertion when the PVA *z*-score is less than −3 result in considerably less flow across the PVA than the Pacifico table criteria. Furthermore, the difficulties of normative scoring are highlighted when we see that a valve designated as having an *z*-score of −3 using one set of *z*-scores can accommodate twice as much flow as another valve designated an *z*-score of −3 using a different set of *z*-scores. Notably, Daubeney-derived *z*-scores [13] result in significantly larger actual PVA diameters for any particular *z*-score. Some reports on TOF repair outcome use *z*-scores that are not readily available for comparison or do not report which *z*-scores were used [14–16]. If these scores correspond to larger actual PVA diameters than the Daubeney *z*-scores, this could explain why some authors report good surgical outcomes when using PVA diameter *z*-scores less than 0 as an indication for TAP insertion. The apparent discrepancy arises because although a small *z*-score (e.g. −2) is quoted by such reports, the actual size of the PVA (in mm) is essentially the same as that corresponding to a *z*-score of 0 or −1 in a report employing Pettersen-derived *z*-scores. The haemodynamic events occurring at the PVA in both instances would be the same; it would then appear as if different TAP criteria have resulted in similar outcomes.

Aggressive pulmonary valve-sparing has been advocated by some [17] due to concerns that TAP insertion increases long-term morbidity and mortality [18]. Interestingly, this has not been found to be the case in a recent large inception cohort study [19]. Conflicting reports such as these complicate surgical decision making. Our data are a step towards quantifying the likelihood of an unacceptable gradient across the PVA in relation to PVA size, thereby allowing surgeons to refine the decision-making process. These data, however, clearly require verification in a larger data set.

LIMITATIONS

This study was performed on a relatively small number of patients from a single institution, therefore the findings may not be generalizable. Higher residual gradients were infrequent. Our institution used the Pacifico table for decision making regarding TAP insertion. As a result, we do not have patients with PVA diameter *z*-scores less than −2. There is a possibility that events occurring at smaller valves could affect the regression line, however in the light of the laws of fluid dynamics, this seems unlikely. Although no patients in our study have required re-intervention for RVOTO, the relatively short follow-up precludes an analysis of re-intervention.

In summary, we are concerned that current TOF repair practice may inappropriately apply PVA diameter *z*-scores to TAP insertion criteria. This practice may result in the preservation of significantly undersized pulmonary valve annuli, leading to unacceptable peak postoperative PVA pressure gradients. Our data, albeit based on a small data set, describe the haemodynamic events occurring at the pulmonary valve in terms of a post-TOF repair PVA pressure gradient in relation to the PVA size described using optimal *z*-scores. We have quantified the likelihood of the occurrence of an unacceptable postoperative gradient across the PVA. We believe that these data could be used to refine the surgical decision-making process and play a role in minimizing the

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number of patients with an unacceptable PVA gradient post-TOF repair; further investigation is warranted.

Conflict of interest: none declared.

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EDITORIAL COMMENT

Repair of tetralogy of Fallot: the right ventricle and the two villains

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Keywords: Congenital heart disease • Tetralogy of Fallot • Right ventricular outflow tract • Pulmonary valve sparing • Ventricular function

In this issue of the Journal, Awori *et al*. [1] propose the criteria for valve-sparing repair of tetralogy of Fallot, with emphasis on the lowest acceptable pulmonary annulus size. It is well known, in practice, that repair of tetralogy of Fallot almost never yields a functional arrangement as good as normal. Even an 'optimal' repair is eventually unsettled by competing failure mechanisms acting more or less independently on the target cardiac chamber, the right ventricle. Due to the anatomical constraints imposed by an hypoplastic pulmonary annulus, the determinants under consideration, i.e. residual right ventricular pressure and/or volume overload (the two villains), are most often only partially tackled at repair and are in fact deliberately maintained in a sort of antagonistic balance. Ideally, repair of tetralogy of Fallot should incorporate a normal-sized and well functioning

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