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Optimal Z-Score Use in Surgical Decision-Making in Pulmonary Atresia With Intact Ventricular Septum

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Abstract

Objectives: In the surgical treatment of pulmonary atresia with intact ventricular septum, the size of the tricuspid valve annulus (as measured by z-scores) has emerged as a significant factor in deciding which repair to perform. Various tricuspid valve annulus z-scores are reported as "cutoffs" for successful biventricular repair. We aimed to determine whether the use of different z-score data sets contributed to the gross variation in "cutoffs" for successful biventricular repair reported in the literature. **Methods:** A single search was made of PubMed using the "advanced" setting with the following search terms: pulmonary, atresia, intact, septum, z, and score. The filters "title" and "title/abstract" were used for the first four and last two terms, respectively; the instruction "AND" combined all terms. Articles that identified which z-score data set was used in patients with biventricular repairs were included. **Results:** From 13 articles, 1,392 patients were studied, 410 (29.5%) of which achieved biventricular repair. Three z-score data sets were quoted; mean tricuspid valve annulus z-scores in biventricular repair patients ranged between −0.53 and −5.1. After correcting for discrepancies between z-score data sets, no study reported a mean tricuspid valve annulus z-score <−2.8 in biventricular repair patients and 83.3% reported mean tricuspid valve annuli z-scores >−1.7. **Conclusion:** The use of varied tricuspid valve annuli z-score data sets may have contributed to gross variations in reported "cutoffs" for successful biventricular repair. This could lead to inappropriate surgical pathway allocation.

Keywords

pulmonary, atresia, z-score

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Introduction

In pulmonary atresia with intact ventricular septum (PA-IVS), the critical anatomical considerations for patient management are the presence/absence of right ventricular/tricuspid valve annular hypoplasia and the presence/absence of a right ventricular–dependent coronary artery circulation (RVDCC).[1,2] The key management decision is whether to perform a biventricular repair (BVR) or a single ventricle repair (SVR); there is evidence that performing a BVR when an SVR is indicated yields a suboptimal outcome.[3,4] The size of the tricuspid valve annulus (TVA), as measured by *z*-scores, has emerged as a significant factor in deciding the repair to be performed.[1] The literature is confusing as various TVA *z*-scores are reported as "cutoffs" for successful BVR.[5,6] A *z*-score of greater than −2 has been quoted as an indicator of successful BVR,[7] however, scores as low as −7.6 have also been reported.[5] It has been shown previously that the indiscriminate use of *z*-scores derived from different populations could lead to different management strategies in similar patients.[8] This study aimed to determine if the use of different *z*-score data sets may have contributed to the gross variation in "cutoffs" for successful BVR in patients with PA-IVS reported in the literature.

Patients and Methods

A single search was made of PubMed using the "advanced" setting. The search terms and filters are shown in Table 1; all filters were combined with the instruction "AND." Retrieved articles were excluded when it was clear from the abstract that

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Table 1. Search Terms and Filters.

Filter	Search Term
Title	Pulmonary
Title	Atresia
Title	Intact
Title	Septum
Title/abstract	Z
Title/abstract	Score

Table 2. TV Z-Score of BVR Patients Versus Type of Z-Score Data Set Used.

Authors	Year	TV Z Score	n (t)	Z-Score Data set
Schneider et al[9]	2014	−1.20 (mn)	24 (60)	Pettersen[10]
Cho et al[11]	2013	−1.07 (mn)	7 (9)	Pettersen
Karamlou et al[12]	2013	−1.00 (mn)	63 (448)	Rowlatt[13]
Chubb et al[14]	2012	−5.10 (mn)	25 (39)	Daubeney[15]
Alwi et al[16]	2011	−2.80 (mn)	17 (143)	Rowlatt
Cleuziou et al[17]	2010	−3.60 (mn)	56 (86)	Daubeney
Alcíbar-Villa et al[18]	2007	−1.10 (mn)	8 (11)	Rowlatt
Daubeney et al[19]	2005	−5.20 (md)	53 (183)	Daubeney
Alwi et al[20]	2005	−1.10 (mn)	44 (53)	Rowlatt
Dyamenahalli et al[21]	2004	−1.65 (mn)	58 (210)	Rowlatt
Minich et al[22]	2000	−0.70 (mn)	23 (36)	Rowlatt
Jahangiri et al[23]	1999	−2.20 (mn)	10 (47)	Rowlatt
Rychik et al[24]	1998	−0.53 (mn)	22 (67)	Rowlatt

Abbreviations: BVR, biventricular repair; md, median; mn, mean; n, number of patients with BVR; t, total number of patients in the study; TV, tricuspid valve.

they were fetal studies. The full text of all remaining articles was examined to determine the *z*-scores of patients who underwent BVR and to determine the *z*-score data set that was used in patients with BVR. Articles that did not specify the data set that was used to calculate the z-scores or which did not examine patients with BVRs were excluded.

Results

Thirty articles where retrieved, 17 of which were excluded. The 13 articles included are listed in Table 2. Of the articles excluded, 7 did not disclose the *z*-score data set used, and the other 10 were excluded for reasons outlined in the Methods section. A total of 1,392 patients were studied and 410 (29.5%) achieved BVR.

One study quoted the median TVA *z*-score and 12 studies quoted mean TVA *z*-scores. After correction for discrepancies between *z*-score data sets (ie, after determining equivalent Rowlatt, Pettersen, and Daubeney *z*-scores; see "Discussion"), none of these 12 had a mean TVA *z*-score <−2.8 and 83.3% (10 of 12) reported mean TVA *z*-scores >−1.7. The discrepancies between data sets were corrected by determining the actual diameter (in mm) of the TVA corresponding to the Daubeney *z*-score for a typical hypothetical patient presented for surgery. This diameter (in mm) was then used to determine the corresponding Rowlatt and Pettersen *z*-scores. In this way, the patient's Daubeney *z*-score was assigned a corresponding (or equivalent) Rowlatt and Pettersen *z*-score.

Discussion

There is a bias with regard to BVR in patients with PA-IVS[12]; patients with hypoplastic right ventricles underwent BVR,[14,17,19] and it has been reported to be possible in patients with TVA *z*-scores as low as −7.6.[5] A recent article[6] revisited some of the important factors considered during surgical decision-making, namely, the size of the TVA as a predictor of BVR and whether small TVAs can be made to grow to a point where they can support a BVR. The authors reported a median TVA *z*-score of −2.79 in their BVR group; this is considerably larger than the −5.2 reported by Daubeney[19] in Table 2. Differences like these in the literature could lead to inappropriate management pathway allocation with resultant suboptimal surgical outcomes.[12] Unfortunately, the *z*-score data set used to calculate *z*-scores was not identified in this recent study.[6] From Table 2, it can be seen that Chubb,[14] Cleuziou,[17] and Daubeney[19]used *z*-scores derived from the same data set[15]; this data set has been shown elsewhere to yield significantly different *z*-scores from the more commonly used Rowlatt data set.[8] For example, the TVA diameter of a neonate with a body surface area (BSA) of 0.2 m^2 and a TVA *z*-score of −5 as described using the Daubeney *z*-score data set[19] is 9 mm. The same neonate would have a TVA *z*-score of −1.3 using the Rowlatt data set.[13] This means that articles using the Daubeney data set (or data sets with similar differences) would report successful BVRs in patients with TVAs as small as −5 described by such *z*-scores, but in actual fact the size of these TVAs (in mm) would be the same as those reported with a *z*-score of −1.3 in articles using the Rowlatt data set. This most likely explains the gross variations in TVA *z*-score "cutoffs" for successful BVR reported in the literature.

The Rowlatt data set was derived from cadaveric studies; the Daubeney data set was derived from echocardiography. Although cardiac structure dimensions may be measured using various modalities, a case for echocardiography has been made and the optimal data set for *z*-score calculation have been proposed[8]; Table 2 contains 2 articles[9,11] that have used this data set. Fortunately, the actual TVA sizes (in millimeters) and the corresponding *z*-scores described by this recommended data set are similar to the Rowlatt data set.[13] Fortunately, because this similarity implies that the inferences obtained from older studies using the Rowlatt data set are transferrable to newer studies using the recommended data set. This is apparent from Table 2 as the mean *z*-score at BVR in the 2 studies using the recommended data set is very similar to that in the older studies that used the Rowlatt data set.

Relatively early on, it was suggested that BVRs might only be successful in patients with TVA *z*-scores larger than −2.5(Rowlatt data used)[25]; a similar recommendation appeared again in the literature more recently.[7] Table 2 seems to support this, as essentially all BVRs were reported from studies where the mean TVA *z*-score was larger than −2.8 (Rowlatt or Pettersen data quoted). What is also apparent from the literature is that small TVAs cannot reliably be made to grow sufficiently to support a BVR and attempts to do so significantly increase operative mortality.[1,6,26] The bias toward BVRs is presumably because the 30-year survival for SVRs[27] is about 40%, and it is anticipated that BVR survival will be better. However, there is evidence that even if a BVR is achieved, right ventricular dysfunction occurs in almost all patients and that BVRs do not ameliorate the damage to the right heart that has been caused antenatally in patients with PA-IVS.[28] Furthermore, outcomes (in terms of exercise capacity) of BVRs achieved in patients with smaller than normal (ie, *z*-score <-3) TVAs are no better than those of SVRs.[12] It has been found that the prevalence of RVDCC increases significantly once the TVA *z*-score drops below -2.[1,23] Attempts to get small TVAs to grow in the presence of an RVDCC greatly increase operative mortality.[1,2] It could be argued that performing BVRs in patients with small TVAs exposes such patients to an increased operative mortality with no outcome benefits. In conclusion, the use of varied TVA *z*-score data sets appears to have contributed to the gross variation in "cutoffs" for successful BVR reported in the literature and could lead to inappropriate surgical pathway allocation. We appreciate that the decision to perform a BVR is not solely based on the diameter of the TVA, however, we feel that when the TVA *z*-score is <-2.8 (as described using the recommended *z*-score data set[8]), the chances of achieving a successful BVR are substantially reduced. While deciding which data set to be used for *z*-score calculation, several factors should be considered: use of recommended measurement technique, types of cardiac structures measured, age range of patients studied, study sample size, use of optimal BSA formula, and quality of images used. The optimal data set for *z*-score calculation was sought from an extensive review of the literature; the Pettersen[10] data set was thought to be optimal.[8] We recommend the use of the Pettersen[10] data set to determine *z*-scores in patients with PA-IVS.

Declaration of Conflicting Interests

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