Surgeons, valve repair in severely unbalanced atrioventricular septal defect, and the emperor's new clothes

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Central Message

Successful valve repair is preferable to replacement. Suboptimal repairs are poorly tolerated in cases of single ventricle, which may warrant reevaluation of the role and timing of valve replacement.

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[I]n short, no one would allow that he could not see these much-admired clothes, because in doing so, he would have declared himself either a simpleton or unfit for his office. Certainly, none of the emperor's various suits had ever made so great an impression as these invisible ones.

"But the emperor has nothing at all on!" said a little child.

"Listen to the voice of innocence!" exclaimed his father; and what the child had said was whispered from one to another.

"But he hasn't got anything on!" the whole town cried out at last. The emperor shivered, for he suspected they were right. But he thought, "This procession has got to go on." So he walked more proudly than ever, as his noblemen held high the train that wasn't there at all.

—"The Emperor's New Clothes,"
Hans Christian Andersen¹

It is a universally acknowledged principle that preservation of native anatomy through successful valve repair is preferable to valve replacement in essentially all clinical settings. The crux of this principle hinges, however, on the word *successful*. Our literature is replete with single-institution observational reports of successful valve repairs of varying types applied to any number of anatomies. There is at least a subliminal, if not frankly overt, message that valve replacement is a form of surgical failure, and the frequency with which valve replacement proves necessary roughly correlates with a surgeon's technical prowess.

The report from Buratto and colleagues² in this month's *Journal* challenges prevailing notions of the role of valve replacement in the management of patients with severely unbalanced atrioventricular septal defect (uAVSD). Perhaps ironically, the article's title invokes the aforementioned traditions of reports of improved outcomes in the setting

of successful valve repair. It is of course true (with certain key caveats such as preserved ventricular function) that if a valve can be repaired with little or no residual insufficiency or stenosis, the patient will fare better than if such an intervention is not undertaken or is unachievable. What is a less obvious and probably underreported statistic is the number of "successful" repairs that carry residual valve dysfunction and in turn increase the ultimate impact of that disease burden. This is a particularly relevant notion in the setting of uAVSD, because repair of a common atrioventricular (AV) valve is a far more challenging undertaking than repair of a tricuspid or mitral valve. (Indeed, the surgical creativity and ingenuity the infinite variety of common AV valve geometry demands make some technical reports on the subject resemble something akin to an National Football League playbook.³) What does the largest series of valve repair in single ventricle forms of uAVSD to date reveal?

- Among patients with uAVSD and single-ventricle anatomy, 1 in 3 required AV valve intervention.
- Of these patients, 1 in 5 did not survive the intervention.
- Nearly half of the cohort did not survive 10 years beyond the intervention.
- When a patient was discharged with moderate or greater AV valve regurgitation, freedom from death or transplantation at 5 years was a dismal 33%, and freedom from re-intervention was 0%.

- Among the small number of patients in the series who underwent AV valve replacement, freedom from death or transplantation at 10 years was a favorable 75%.

These are sobering data from a series purporting to describe improved outcomes in patients with successful valve repair. The report possesses the usual weaknesses of retrospective single-institution studies that span forty years. Among these, it is important to note the relative paucity of information about the role of ventricular dysfunction in both valve repair efficacy and overall clinical outcomes. Buratto and colleagues² did not find a correlation between ventricular dysfunction and outcome, but the data are merely descriptive and were only available for 33 of the 44 patients in the series. The role of ventricular dysfunction in the outcome of these patients is well documented⁴ and powerful, and a more granular analysis of ventricular function in this cohort would have been helpful.

What is refreshing about this manuscript, however, is the objective and unapologetic analysis by Buratto and colleagues² of the results and their implications. The message from these data is clear: these patients are

ill-served by strategies that equate success with tolerance of residual disease. Buratto and colleagues' suggestion² that valve replacement may best be applied earlier in the clinical sequence of valve interventions represents a nontrivial departure from the current wisdom. And lest we become like the emperor's sycophants, our collective comfort with "successful" valve repairs in this patient population should perhaps lessen, and a fresh assessment of the role of traditional mechanical valve replacement be entertained.

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