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Editorial

Pulmonary atresia with intact ventricular septum—a continuing challenge for both cardiologist and surgeon

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With the exception of its counterpart involving the left side of the heart, our lack of success in modifying the dismal natural history of the hypoplastic right heart syndrome represents one of the most obvious failures of modern pediatric cardiology and surgery. Published results vary, but by-and-large, the neonate with pulmonary atresia and intact ventricular septum has considerably less than a 50% chance of surviving into adulthood with a fully corrected biventricular heart.[1-3] Despite an ever increasing body of literature, how far have we progressed after more than 20 years of active management of these patients? In this issue, the fetal cardiology group from Guy's Hospital, London, specializing in perinatal cardiology, present their data from fetuses which extends our understanding of the natural history of this condition (page 367), while at the other end of the temporal spectrum, the cardiopulmonary physiology of the fully corrected heart is explored (page 382). We also have the opportunity to examine the long-term institutional results from the Royal Brompton Hospital and the Royal Liverpool Hospitals (pages 377 and 395), as well as the more contemporaneous results of a similar surgical protocol adopted by Vosa and coworkers in Naples (page 391). Inevitably, there still remain more questions unanswered than answered but, in this brief review, we will attempt to highlight some of the most important issues regarding the management of this most challenging group of patients.

Preoperative diagnosis—the coronary arteries

It could be argued that, in a situation where the management of these patients is so clearly in a state of development, the cardiologist is obliged to obtain as much anatomical and physiological data as possible prior to embarking on a particular protocol for management. To this end, many perform cardiac catheterization with angiography, in addition to diagnostic cross-sectional echocardiography, in all neonates presenting with pulmonary atresia and with an intact ventricular septum. If one takes a slightly more pragmatic approach, however, the value of cardiac catheterization may be challenged. Assuming adequate cross-sectional echocardiography is available, then the role of cardiac catheterization in the neonate is limited to the demonstration of the coronary circulation. There can be no doubt that "important" abnormalities of coronary perfusion exist in a significant proportion of patients.[1] But, other than when there is an anomalous origin of a coronary artery from the pulmonary trunk (a situation encountered infrequently), how does this information influence outcome after initial palliation? Cardiac catheterization was not performed in the neonatal period, but the results of primary palliation of all patients by insertion of a Blalock-Taussig shunt, as reported in this issue by Vosa and colleagues,[4] are second to none (one death in 41 consecutive patients). It is difficult to argue that cardiac catheterization would have improved the early outcome in their patients, particularly when neonatal transplantation did not exist as an alternative for those with "inoperable" coronary lesions. The situation is quite different, however, if early decompression of the right ventricle is contemplated. The Toronto group has emphasized the importance of the right ventricular dependency of all or, more commonly, part of the coronary arterial blood supply.[5] Severe stenosis or interruption of the native coronary arteries may be present at birth or may develop with time. Decompression of the right ventricle under these circumstances may lead to myocardial ischemia, infarction, and death.[6] Heroic attempts to secure myocardial perfusion have been made by grafting the coronary arteries but, for some neonates, early cardiac transplantation is the only viable option. Whether it is possible to influence the progression of sinusoid-related coronary arterial occlusion is open to debate. Right ventricular thrombo-occlusion, while intellectually attractive, is associated with a significant mortality, and its long-term utility remains unproven.[7] Nonetheless, one must accept the funda-

mental importance of adequate myocardial perfusion under all circumstances. Indeed, there is circumstantial evidence to suggest that the ventricular dysfunction which occurs as a consequence of ventriculocoronary connections is of fundamental importance to ultimate outcome.[8,9] Thus, the notion that the management of this condition must conceptually include, and be guided by, a thorough knowledge of the coronary arterial anatomy seems unchallengeable, even though the practical benefits of this knowledge are yet to be convincingly demonstrated.

Surgical protocols—the right ventricle

If one analyses the results of long-term institutional experience of this disease, one is struck by the many and varied surgical protocols that have been adopted. This is related to many factors. We can highlight a few; first, the diversity of the anatomic substrate, from "simple" valvar atresia (which may be corrected in a single stage) to extreme right ventricular hypoplasia with major abnormalities of the coronary arteries; second, the relatively poor outcome for the group as a whole, almost no matter what protocol is adopted and how often it is changed; third, and, perhaps most importantly, a refinement of approach which is based on experience and improved understanding of the disease. It is impossible to pigeon-hole these patients into rigid protocols because, to some extent, the management of each patient with pulmonary atresia has to be individualized. Nonetheless, there are some basic principles which should guide their treatment. While the theoretical importance of the coronary arterial anatomy has been discussed, the practical importance of right ventricular size cannot be overemphasized. In those with a diminutive ventricle, the protocol is straightforward: preparation for an atriopulmonary or cavopulmonary circulation. At the other end of the spectrum, there will be a small group of patients in whom the right ventricle has dimensions close to normal. These should be suitable for a biventricular correction in a single stage.[8] For the majority of patients, the decision is less clear-cut. A multi-stage protocol is inevitable, and most now include a modified Blalock-Taussig or central aortopulmonary shunt with coincident or later reconstruction of the right ventricular outflow tract, in order to enhance right ventricular growth. The exact details and results of this approach vary from institution to institution (as is amply demonstrated in the surgical studies published in this issue). Their relative merits will not be discussed further. Much has been written, nonetheless, in regard to the assessment of right ventricular dimensions, growth and subsequent suitability for a biventricular correction in this group of patients.[9-11] Right ventricular size will increase in the majority after reconstruction of the right ventricular outflow tract, but it is important to remember that right ventricular dilatation resulting from an increased stroke volume can occur in one of two ways, usefully and uselessly. In the former, an increased antegrade preload (via a non-limiting inflow to the right ventricle) leads to a usefully enlarged right ventricle and, ultimately, suitability for biventricular repair. In the latter, useless right ventricular dilatation may result from a "retrograde" preload in response to pulmonary regurgitation. While the right ventricular cavity may enlarge under these circumstances, a restrictive right ventricular inflow may make biventricular repair impossible. This further emphasizes that surgical decision-making, and subsequent analysis of results, can only sensibly be performed by taking account of all three constituent parts of the 'tripartite' right ventricle.[12]

Definitive biventricular repair

Assuming that the right ventricular dimensions are adequate, then biventricular repair remains the ultimate therapeutic goal for patients with pulmonary atresia and intact ventricular septum. In a recently published editorial, Freedom asked the rhetorical question, "How can something so small cause so much grief?"[13] As we have discussed above, right ventricular size, in all three of its components, is fundamental to outcome following biventricular repair. Right ventricular systolic performance is rarely at issue in these hearts, the stroke volume being limited by its end-diastolic, not its end-systolic, volume. It would seem, therefore, that the diastolic properties of the right ventricular myocardium would be the most important in determining cardiac output and, hence, survival in the immediate postoperative period. Nonetheless, the information available regarding cardiopulmonary physiology either before or after biventricular repair is remarkably scarce.[14] On page 382 of this issue, our own data regarding the diastolic properties of the right ventricle in patients after biventricular repair are presented.[15] It would appear that, even in those patients who have successfully undergone fully "corrective" surgery, the right ventricle has a fixed end-diastolic volume. In other words, it is restrictive. This is manifest both in terms of the characteristics of its early diastolic filling and its late diastolic function as a conduit, atrial systole merely pushing blood through the ventricle rather than filling it. The effects of normal respiratory augmentation of cardiac output, and the normal beneficial effect of atrial systole on cardiac output, are both more pronounced in these patients. Do these findings have any practical significance? There seems little doubt that, in the immediate postoperative period, the maintenance of sinus rhythm and the minimalization of mean airway pres-

sure during positive pressure ventilation will be important in optimizing cardiac output. If the right ventricle is borderline, or is simply too small to support the entire cardiac output, then other maneuvers have been proposed. Fenestration of the atrial septum, with subsequent closure, has been used by Laks and coworkers to good effect.[16] Thus, in the early post-operative period, systemic cardiac output is maintained at the expense of arterial desaturation, in the same way as in the fenestrated Fontan circulation. Another strategy is to reduce the volume load on the right ventricle. Biventricular repair, but with prior bidirectional superior cavopulmonary anastomosis, will lead to a physiological "correction," but with the right ventricle only having to impart energy to two-thirds of the systemic venous return. It is, of course, impossible to make definitive statements regarding either the physiological consequences or clinical utility of these various surgical approaches, as the numbers treated thus far are inevitably so small. What is clear, however, is that "all right ventricles in this condition are restrictive, but some are more restrictive than others!"

Non-surgical correction of pulmonary atresia with intact ventricular septum

The ever-widening indications for interventional catheterization in pediatric cardiology now include the treatment of pulmonary atresia with intact ventricular septum. This approach also holds good for critical pulmonary stenosis. It is salutary to note that surgical results are almost equally as dismal for critical valvar stenosis presenting in the neonatal period as they are for valvar pulmonary atresia. The congenital heart surgeons study, and the data from the Royal Brompton Hospital (see page 377 of this issue), suggest a similar overall mortality. This is despite the infrequency with which coronary arterial abnormalities are seen in patients with critical valvar pulmonary stenosis. We come back, perhaps, to the problem of the restrictive right ventricle. Non-surgical treatment under these circumstances should, and does, confer a considerable advantage. Indeed, balloon dilatation of critical pulmonary valvar stenosis is now routinely performed in many institutions. But for a pinhole orifice in the pulmonary valve, some patients with pulmonary atresia and intact ventricular septum are indistinguishable in terms of both physiology and anatomy (and, as intimated, perhaps even outcome after surgery). In the past year or so, Qureshi and his colleagues from Guy's Hospital, London, have described a technique whereby patients with imperforate pulmonary valves are "converted" into critical pulmonary stenosis by laser-assisted perforation of the valvar membrane.[17] Although applicable only to those patients with an imperforate valve, the results are little short of sensational. Non-surgical correction, with discharge from hospital within a few days of birth, can now be achieved. We are still learning with this technique. It is not without its own complications and the learning curve includes significant morbidity and mortality.[17] With increasing experience and refinements in the technique (see page 387 of this issue)[18] these problems should be reduced. Its precise role remains to be established, but the technique is surely here to stay.

Conclusion

While there is still a lot to learn, the importance of the coronary arterial anatomy and right ventricular morphology on subsequent outcome is becoming increasingly understood. Whether theory in practice will lead to improved results has yet to be demonstrated. At present, we can only conclude that the patient with pulmonary atresia and an intact ventricular septum remains a challenge to cardiologist and surgeon alike.

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