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Editorial

The potentially obstructive subaortic region and banding of the pulmonary trunk—selected observations in the patient considered for a Fontan procedure

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Summary There is a clear and present danger for the patient considered for Fontan's operation in the presence of excessive ventricular hypertrophy and/or mass. Such ventricular hypertrophy and/or mass probably reflects singly, or in combination, banding of the pulmonary trunk and subaortic stenosis, as well as other factors. While one can construct a number of therapeutic algorithms for patients seen at various ages with a restrictive ventricular septal defect and subaortic stenosis in the setting of double inlet left ventricle and discordant ventriculoarterial connections, the optimal management should be to minimize the potential risks for development of subaortic stenosis and excessive ventricular hypertrophy and/or mass.

Key words: Univentricular atrioventricular connections; double inlet left ventricle; tricuspid atresia; banding of the pulmonary trunk

Of the original criteria of Choussat and his colleagues,[1] normal diastolic function was listed as one of the so-called 'ten commandments' necessary for the successful performance of a Fontan operation. Yet, and perhaps somewhat surprisingly, in the more than two decades since Fontan and Baudet[2] performed their now classic surgery, the evaluation of diastolic function in patients with complexly malformed hearts being considered for Fontan's operation or one of its many variations still remains controversial. And, while considerably more attention has been focused on systolic function, the methodology used even in this arena is likely to stimulate considerable discussion.

The specific relationship in these complex hearts between myocardial hypertrophy and diastolic function or compliance is unclear. What has become evident is that myocardial hypertrophy is a risk factor for the function of a heart corrected by a Fontan procedure.[3-7] For patients with excessive ventricular hypertrophy prior to the operation, the construction of an atriopulmonary connection and atrial separation will decrease the ventricular volume while the ventricular mass seemingly increases, perhaps altering ventricular compliance. Such patients do not fare well at Fontan's operation, and their frequent subsequent course leading to death, takedown of the procedure, or increased morbidity supports this observation.[3,4,6-10]

Penny and his colleagues[11] have studied the acute transition to a Fontan circulation. Their data from 10 patients undergoing a transition to a Fontan state indicates that there is acute ventricular hypertrophy reflecting a sudden reduction in ventricular preload. Ventricular compliance was unchanged in their patients, but the time constant of ventricular relaxation was prolonged and, thus, early diastolic filling velocity was reduced.

Etiology of myocardial hypertrophy

We, and others, have identified at least two major causes of abnormal myocardial hypertrophy in patients considered for the Fontan operation, namely subaortic stenosis and banding of the pulmonary trunk. There is considerable data that these two risk factors are often identified in the same patient and, thus, banding of the

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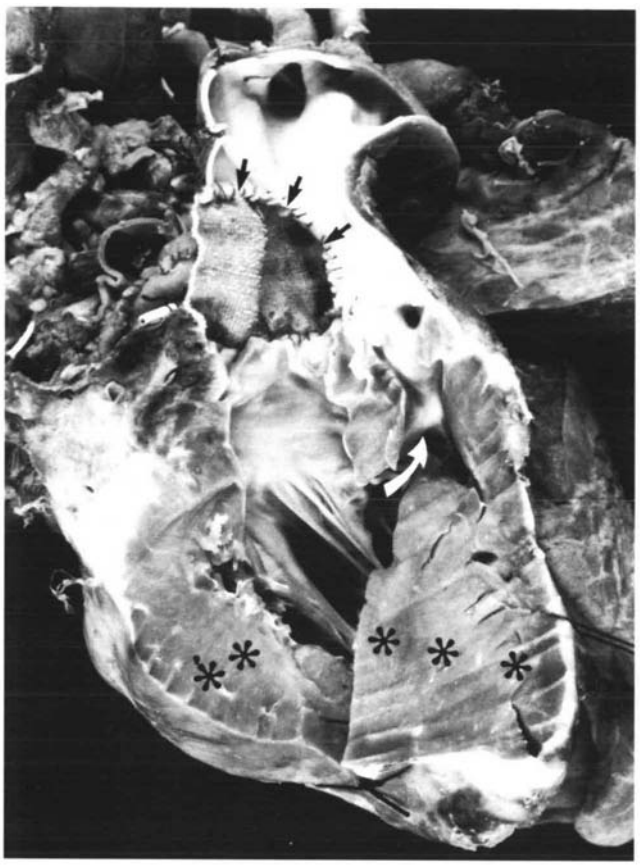

Figure 1. *Massive ventricular hypertrophy(*) in a patient dying after the Fontan and Damus-Kaye-Stansel procedures (black arrows). The ventricular septal defect is restrictive (white arrow).*

pulmonary trunk results in or is causal of subaortic stenosis. This view is not universally held, and the reasons for this divergence of opinions will be discussed later.[12-14]

If abnormal myocardial hypertrophy translates into poor results after the Fontan procedure, what are the types of cardiac malformation where such myocardial hypertrophy might be identified, and what predisposes these hearts to abnormal ventricular hypertrophy?

Table 1. Volumetric and wall mass data from patients with good outcomes.

Parameter	Pre-Fontan (n=23)	Post-Fontan (n=31)	Control (n=25)
Age	6.1 ± 3.5	8.4 ± 3.6	4.1 ± 2.2
EDVI(ml/m²)	99 ± 23	63 ± 12	64 ± 14
ESVI(ml/m²)	49 ± 16	31 ± 9	26 ± 8
WMI(g/m²)	106 ± 33	75 ± 22	63 ± 16
WMI/EDVI(g/ml)	1.08 ± 0.31	1.20 ± 0.38	0.97 ± 0.1

EDVI: end-diastolic volume index; ESVI: end-systolic volume index; WMI: left ventricular wall mass index.

Dominant left ventricle with rudimentary right ventricle and discordant ventriculoarterial connections

This is not the appropriate forum to discuss all those malformations potentially suitable for construction of a Fontan circuit. At the Hospital for Sick Children in Toronto, in patients considered for Fontan's operation, the most common anatomical substrate for subaortic stenosis is the heart characterized by a dominant left ventricle, a rudimentary right ventricle, and discordant ventriculoarterial connections. The communication between the dominant left ventricle and rudimentary right ventricle has been termed the ventricular septal defect or bulboventricular foramen, although there is not unanimity about this designation.[15] While initially using, and indeed advocating, this latter term, we now prefer a morphological designation, rather than an embryological one. Thus, tricuspid atresia or double inlet left ventricle with discordant ventriculoarterial connections are common forms of congenitally malformed hearts where subaortic stenosis might develop, especially when there is not important naturally-occurring pulmonary outflow tract obstruction. When restrictive, the ventricular septal defect in this setting results in subaortic stenosis. In addition to restriction at the septal defect, obstruction within the rudimentary right ventricle below the aortic root may also contribute to subaortic stenosis.[16] The presence of important obstruction of the subpulmonary outflow tract in such patients does not preclude the possibility of significant subaortic stenosis.[9,17,18] Some views about the nomenclature of these hearts have recently been summarized in a provocative editorial.[19]

The ventricular septal defect in double inlet left ventricle

Anderson and his colleagues[20] have thoroughly reviewed the variable morphology of the ventricular septal defect in hearts with double inlet left ventricle. For the most part, among those hearts with discordant ventriculoarterial connections, the ventricular septal defect is muscular, as the membranous septum has not been formed. In some of these patients the aortic valve roofs the defect, but even in this situation it is still a muscular defect.[17] The shape of these defects is usually not circular, but more elliptical, like a 'button-hole,' and for this reason, imaging may suggest the defect is of adequate size in one plane, while in the other it is not.[18,21,22] The perception of the defect as elliptical has

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been used clinically from echocardiographic parameters to predict which holes might become obstructive.[23]

Double inlet left ventricle, discordant ventriculo-arterial connections, and an intact ventricular septum

The ventricular septal defect in a rare patient with a double inlet left ventricle or tricuspid atresia and discordant ventriculoarterial connections may be intact.[16,24-29] Systemic blood flow in these patients is thus dependent on flow through an arterial duct, but unlike the patient with the usual form of hypoplasia of the left heart, an obstructive interatrial septum does not contribute to the potential difficulties. The therapeutic options for patients with these uncommon situations includes either a staged Norwood approach concluding in a Fontan operation or cardiac replacement. Because the aortic valve is often abnormal in these patients, a palliative arterial switch would be unlikely to prove successful in these patients.

Double inlet left ventricle, discordant ventriculo-arterial connections, and clearly restrictive ventricular septal defect (and thus subaortic stenosis)

The ventricular septal defect in some hearts with double inlet left ventricle and discordant ventriculoarterial connections is unequivocally and to all standards obstructive. With very rare exception in these patients, there is no evidence of fixed obstruction to the pulmonary outflow tract, and clinical symptomatology reflects a torrential pulmonary blood flow, pulmonary arterial hypertension, and the frequently present obstructive anomaly of the aortic arch, including coarctation, aortic arch atresia, or even interruption.[17,21,30] Because of the parallel circulations, and the often profound degree of congestive heart failure observed in these situations, a pressure gradient between dominant left ventricle and aorta is infrequently observed and recorded. Yet with cross-sectional echocardiographic imaging, Doppler and color-flow interrogation, such septal defects are unequivocally smaller than the aortic root, often 50% or less, and one would be most concerned about their fate[17,31] (Figure 2). We,[8,10,18] like Franklin and his colleagues,[30] have taken a most conservative posture in considering the role of banding of the pulmonary trunk as effective palliation. Indeed, we are persuaded that banding should be eschewed in these young patients, and there are now a number of alternatives that can protect both the myocardium and the integrity of the pulmonary vascular bed for a later Fontan procedure (Figure 3). Such alternatives in the neonate and the young infant include a Norwood-like approach with pulmonary blood flow mediated through a systemic-to-pulmonary artery connection, the palliative arterial switch procedure as carried out by Karl and his associates,[32] or surgical enlargement of the septal defect.[5,33] We have recently summarized these surgical alternatives.[9]

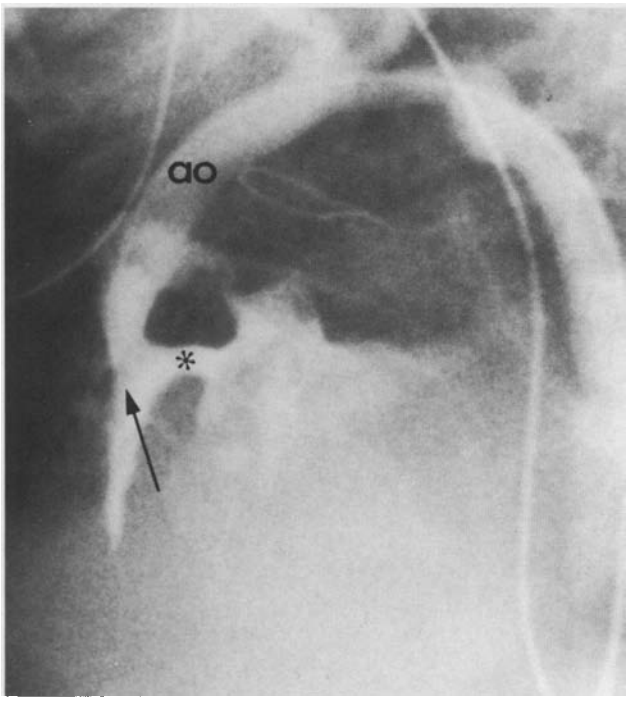

Figure 2. *A severely restrictive ventricular septal defect(*) in a neonate studied angiographically within days of repair of a coarctation of the aorta and banding of the pulmonary trunk. The rudimentary right ventricle (black arrow) is slit-like. Left ventricular angiogram in long axial oblique projection. ao: aorta.*

The hemodynamic definition of subaortic stenosis in the patient with double inlet left ventricle and discordant ventriculoarterial connection—What is the essence of subaortic stenosis?

We have defined earlier the morphological substrate for subaortic stenosis in the patient with double inlet left ventricle and discordant ventriculoarterial connection—namely, a restrictive ventricular septal defect with or without narrowing of the subaortic infundibulum. The definition of the anatomic substrate poses little controversy. The hemodynamic recognition of subaortic stenosis in such hearts with pulmonary hypertension or a band around the pulmonary trunk remains controversial. Stated simply, what is the essence of the diagnosis of subaortic stenosis in these patients? In the symptomatic neonate and young infant prior to palliative therapy, the absence of pressure gradient, even a minor one, does not exclude important subaortic stenosis, as the left ventricle can eject blood into both the unobstructed

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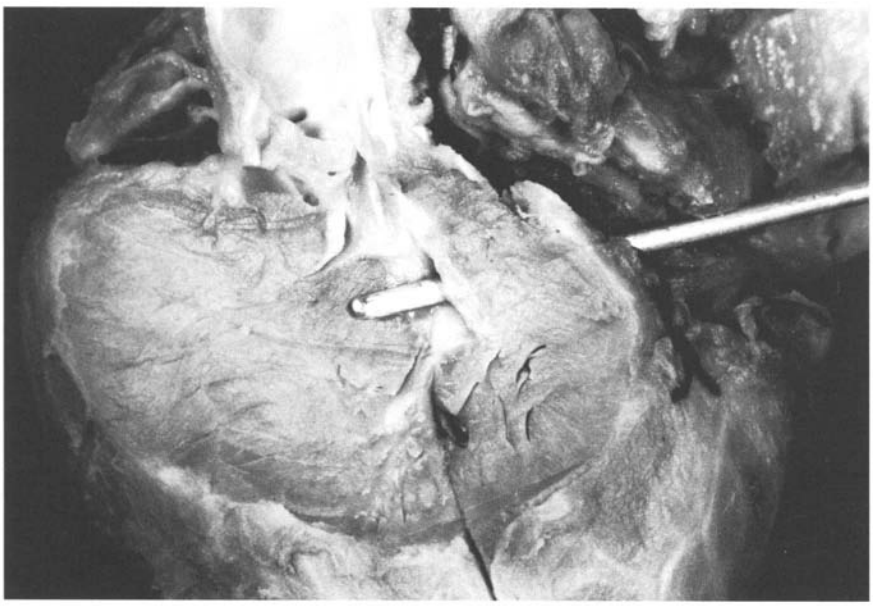

Figure 3. *Massive ventricular hypertrophy in a neonate with a dominant left ventricle, rudimentary right ventricle and discordant ventriculoarterial connections.*

pulmonary arterial bed as well as the aorta. We have shown elsewhere that the ventricular septal defect may be very small, but as long as there are parallel outlets, despite an adequately banded pulmonary trunk, a pressure gradient between aorta and dominant left ventricle may not be present.[16,18] Others still rely on some pressure gradient before unequivocally making the diagnosis of subaortic stenosis. It has been our contention for a number of years that, by the time any pressure gradient is evident, subaortic stenosis is well-established, reflecting a significantly obstructive defect. In the absence of a resting pressure gradient, isoprenaline may well provoke a systolic gradient, an observation reported by Somerville and her colleagues in 1974,[34] and subsequently advocated as a useful maneuver from our institution.[16,18,21] If one cannot rely on the presence of a systolic pressure gradient to establish the diagnosis of subaortic stenosis, then the essence of subaortic stenosis must be the character of the ventricular septal defect. Then one must ask—How small is too small? It has been our position that when the ventricular septal defect size is smaller than that of the aortic orifice, the defect is restrictive, a view held by Rychik and his colleagues at the Children's Hospital of Philadelphia.[35] But what should the exact ratio be: 75%, 50%, etc.? In a patient whose defect is unequivocally smaller than the aortic root, banding of the pulmonary trunk will also alter the wall mass index/end-diastolic volume index, and thus subaortic stenosis is an unfortunate inevitability, reflecting both the dynamic changes in cardiac form and function and the natural history phenomenon (Figure 4). For a fortunate few patients requiring banding, the septal defect will not be restrictive, or minimally so, at least initially.

The role of banding in the palliation of double inlet left ventricle and discordant ventriculoarterial connections with no pulmonary outflow tract obstruction

Following the clinical observations of Somerville and her colleagues[34] who described two children with double

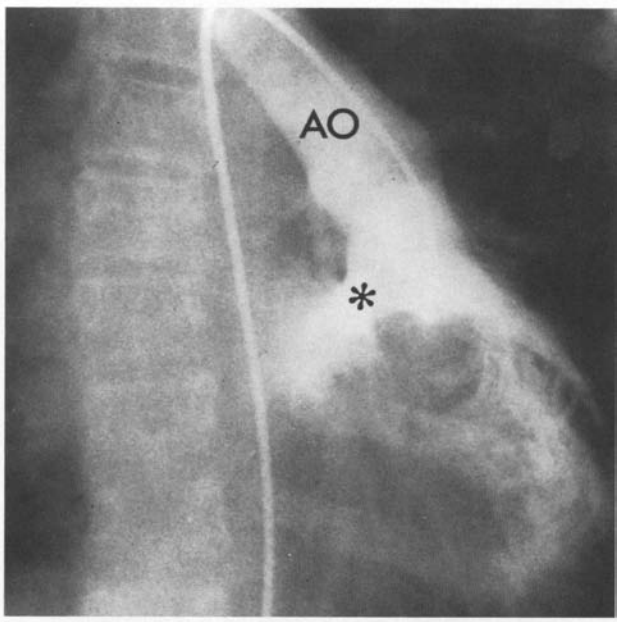

Figure 4. *Restrictive outlet ventricular septal defect(*) in a patient with double inlet left ventricle, discordant ventriculoarterial connections and multiple smaller trabecular ventricular septal defects. The larger outlet ventricular septal defect(*) is certainly smaller than the aortic root (AO) in this projection.*

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inlet left ventricle and discordant ventriculoarterial connections, both of whom acquired important subaortic stenosis after palliative banding of the pulmonary trunk (although these authors did not suggest a causal relationship between the banding and subaortic stenosis), we were persuaded that subaortic stenosis was a sequel of banding.[36] In a series of papers emanating from this institution, we explored this association.[8-10,18,21,37] Initially, we advocated the view that banding was possibly causal of subaortic stenosis. This view is certainly not shared by Rao.[12-14] It is his suggestion that the ventricular septal defects in patients subjected to banding are those that tend to spontaneous diminution in size, and that banding, by improving patient survival, probably allows patients to express the natural history of the initially restrictive defect. In this regard, Franklin and his colleagues[38,39] have shown that survival before definitive repair was particularly poor in those patients with double inlet left ventricle, discordant ventriculoarterial connections, systemic arterial obstruction, and high pulmonary blood flow. We also share the view that the ventricular septal defects in patients with no obstruction to pulmonary blood flow in the setting of double inlet left ventricle and discordant ventriculoarterial connection tend to be smaller than the aortic root than in those patients with similar morphology and naturally-occurring important subpulmonary obstruction,[8,10] observations made some years later by Bevilacqua and associates.[17] We would emphasize that banding of the pulmonary trunk by promoting muscular hypertrophy probably accelerates the tendency of these muscular defects to spontaneous diminution in size.[8-10] Thus, no matter what the specific relationship between banding and the development or unmasking of latent subaortic stenosis, both events contribute to myocardial hypertrophy. And myocardial hypertrophy impacts in an unfavorable way on outcome after the Fontan procedure.[3-10] In this regard, Franklin and his associates[26] reviewed the outcome and potential for definitive repair of patients with double inlet ventricle presenting in infancy. They found, and not surprisingly, that patients requiring no initial palliative operation and those undergoing a systemic-to-pulmonary arterial shunt, fared better than those who, first, underwent isolated banding of the pulmonary trunk or, second, required repair of the aortic arch and banding of the pulmonary trunk, the latter because of the development of subaortic stenosis.

Thus, is there a role for banding of the pulmonary trunk in any patient in whom the Fontan procedure is contemplated? Our unit is most cautious in the use of banding of the pulmonary trunk for the reasons articulated above. We would not advocate banding when double inlet left ventricle and discordant ventriculoarterial connections is found, first, with an intact or nearly intact ventricular septum, second, with a ventricular septal defect and obstructive anomaly of the aortic arch, and, third, when the ventricular septal defect is restrictive.

Not all patients with double inlet left ventricle and discordant ventriculoarterial connections who are subjected to banding of the pulmonary trunk develop subaortic stenosis,[30] although the majority of patients reported from Toronto do so.[10] For some patients with a slightly small defect, it seems reasonable to band the pulmonary trunk for a few months until a Damus-Kaye-Stansel and bidirectional cavopulmonary connection can be constructed. Additionally a minority of patients with double inlet left ventricle, discordant ventriculoarterial connections, torrential pulmonary blood flow and pulmonary hypertension can be managed with banding without developing subaortic stenosis, with their surgical management concluding in a favorable Fontan outcome.[30]

But there is persuasive data from a number of institutions that the Fontan procedure is palliative at best,[4,40,41] with late survival to 10-15 years after Fontan of only 50-70%. This caveat is applicable to patients with tricuspid atresia, but even more so to those with double inlet left ventricle and discordant ventriculoarterial connections.[40] For those patients who undergo banding of the pulmonary trunk and any of a series of other surgical maneuvers eventually concluding in Fontan's operation, is the long-term history of these patients similar to those whose surgical history prior to the Fontan does not include banding? What is the long-term effect of banding on the neonatal and infant myocardium in the setting of double inlet left ventricle? Data from another group of patients[42] suggests that the left ventricular myocardial mass may increase rapidly after pulmonary banding, and this observation set the stage for a rapid two-stage arterial switch conducted beyond the neonatal period. But are such changes reversible, or do they intimate something about the future for these patients?

In summary, banding of the pulmonary trunk is not a realistic therapeutic procedure for many patients with double inlet left ventricle and discordant ventriculoarterial connections. The inevitability of myocardial hypertrophy in patients subjected to banding, the reality of subaortic stenosis(especially in those patients with pre-existing obstructive anomalies of the aortic arch) and concerns about the survival of patients previously banded after Fontan's operation all urge caution in the application of this once standard technique.[8]

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