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ORIGINAL ARTICLE

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Clinical findings in right ventricular noncompaction in hypoplastic left heart syndrome

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Abstract

Background: Noncompaction is a poorly understood form of cardiomyopathy that typically affects the left ventricle and may be associated with congenital heart disease. Right ventricular noncompaction (RVNC) may occur when the left ventricle is affected but is rarely seen in isolation. RVNC may have clinical significance affecting surgical and long-term outcomes. We describe the diagnosis and clinical course in three patients at our institution.

Methods: We performed a retrospective review of patients diagnosed with RVNC over a 12-month period at our institution and reviewed their imaging and clinical course.

Results: Three patients were identified. All had diagnosis of RVNC by echocardiography (echo) made on postnatal imaging which reviewed degree of trabeculation, and noncompaction-to-compaction ratio of the myocardium. Patient A was a neonate with hypoplastic left heart syndrome (HLHS) who underwent a Norwood operation with Sano modification. Her postoperative course was notable for low-normal RV function. She returned with a pericardial effusion warranting immediate pericardiocentesis. She continued to have effusions, which were medically managed. She was subsequently found to have an RV apical pseudoaneurysm, which required surgical resection. Patient B was a neonate with HLHS who had a Norwood operation with Sano modification. She had low-normal RV function on echo. She required medical management for pericardial effusion. Patient C was a neonate with HLHS who also underwent a Norwood operation with Sano modification. His postoperative course was notable for elevated serum brain natriuretic peptide, which was treated with digoxin.

Conclusion: RVNC is a rare diagnosis with limited known clinical impact. One of these patients had a very rare complication after pericardiocentesis (pseudoaneurysm) that may have been related to the RVNC. Our understanding of this disease process is limited and requires additional investigation, but emphasizes the importance of appropriate diagnosis to allow for timely follow-up and counseling for this unique population.

KEYWORDS
clinical outcomes, hypoplastic left heart syndrome, noncompaction cardiomyopathy

1 | INTRODUCTION

Left ventricular noncompaction (LVNC) is a poorly understood form of cardiomyopathy that typically affects the left ventricle and may be seen in isolation or in association with congenital heart disease.[1–3] Right ventricular noncompaction (RVNC) may occur when the left ventricle is affected but has only rarely been reported in isolation. We present a series of three patients diagnosed with isolated RVNC in the setting of hypoplastic left heart syndrome (HLHS). Though there are no echocardiographic criteria for RVNC, we applied the criteria used in LVNC. Thus, diagnosis was made by postnatal assessment of the degree of trabeculations, and the noncompaction-to-compaction ratio of the myocardium of at least 2:1 (Figure 1).[4,5]

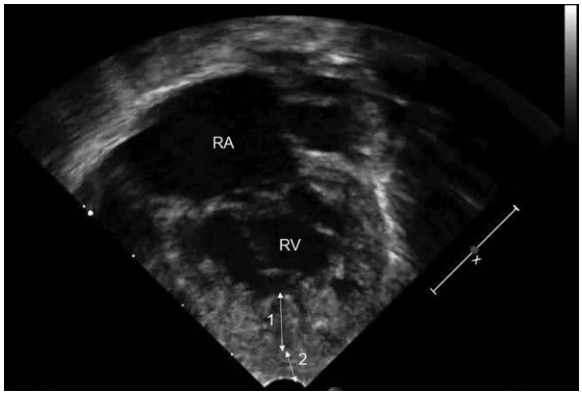

FIGURE 1 Echocardiogram for patient B: apical view demonstrating HLHS with heavily trabeculated right ventricle (RV). The noncompacted myocardium (measurement 1) compared to the compacted myocardium (measurement 2) is more than 2:1, consistent with noncompaction. RA = right atrium

2 | PATIENT A

A neonate with a prenatal diagnosis of HLHS underwent a Norwood operation with Sano modification within the first week of life. Her postoperative course was notable for low-normal right ventricular function at the time of stage I discharge, with postoperative echocardiography demonstrating hypertrabeculation of the RV consistent with RVNC. She presented within 3 weeks of initial hospital discharge with a large pericardial effusion warranting immediate pericardiocentesis. She continued to have pericardial effusions, which were followed on serial echocardiography and managed with diuretics and oral corticosteroids. She was subsequently found to have a pseudoaneurysm at the right ventricular apex, which required surgical resection at 6 weeks of age (Figures 2 and 3). At the time of catheterization prior to her stage II operation, she was found to have recoarctation of her aorta which required balloon angioplasty. Her stage II (Glenn) operation occurred at 5 months of age without immediate complication. Due to mild to moderate RV dysfunction after Glenn, digoxin was started. Function normalized in the months following stage II discharge.

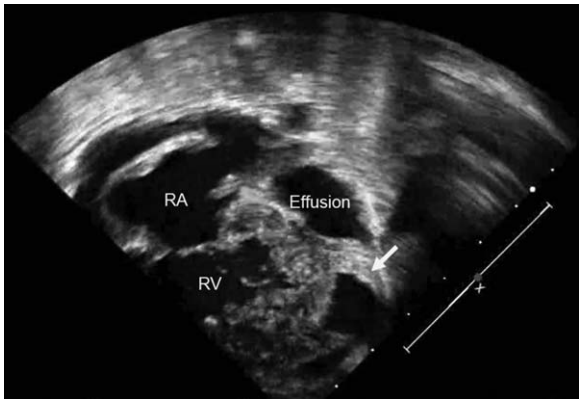

FIGURE 2 Echocardiogram for patient A: apical view demonstrating HLHS with dilated and noncompacted right ventricle (RV). Also seen is a moderate global pericardial effusion as well as an apical pseudoaneurysm (arrow). RA = right atrium

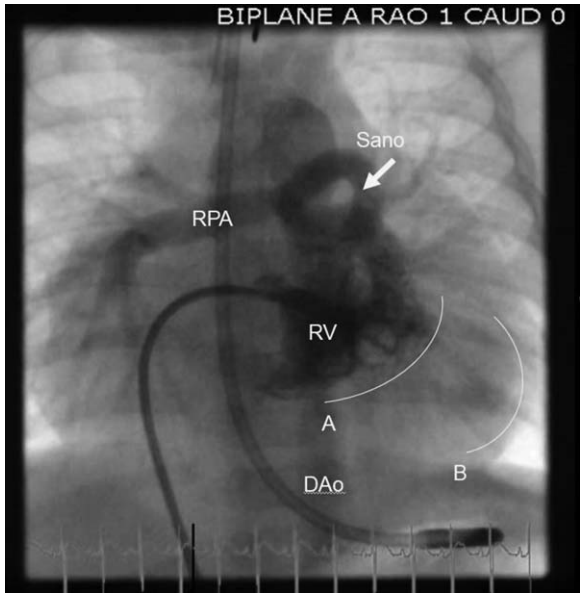

FIGURE 3 Angiography for patient A: Power injection via catheter in right ventricle (RV) (outlined A) with pseudoaneurysm (outlined B). Also seen are the heavy trabeculations of the RV. RPA = right pulmonary artery, Dao = descending aorta

3 | PATIENT B

A neonate with prenatal diagnosis of HLHS and immediate postnatal echocardiogram with findings of RVNC had a Norwood operation with Sano modification, as well as unroofing of the coronary sinus at 3 days of life. Postoperative right ventricular function was qualitatively low-normal, with a moderate pericardial effusion that was managed with steroids, but did not require intervention. Prior to stage II operation, she was found to have recoarctation of the aorta which required catheter-based intervention. Prior to her stage II operation, she was placed on milrinone infusion for poor right ventricular function. She underwent stage II operation at 5 months of age which consisted of a hemi-Fontan operation and pulmonary arterioplasty. Due to persistent right ventricular dysfunction postoperatively, she was transitioned from milrinone to oral digoxin and enalapril for discharge. During her stage II operation admission, she also had persistent chylous pericardial and pleural effusions, both of which required drainage procedures. She has subsequently undergone further evaluation for abnormal lymphatic circulation with catheter-based intervention.

4 | PATIENT C

A neonate with prenatal diagnosis of HLHS and immediate postnatal echocardiogram demonstrating RVNC with restrictive atrial septum underwent Norwood operation with Sano modification and atrial septectomy at 7 days of age with no immediate complication. He had normal right ventricular function after surgery, but due to elevated

TABLE 1 Summary of subject outcomes

	Patient A	Patient B	Patient C
Cardiac anatomy	HLHS (MA/AA)	HLHS (MA/AA)	HLHS (MA/AA)
Stage I operation	Norwood with Sano modification	Norwood with Sano modification	Norwood with Sano modification, atrial septectomy
Stage I complications	Pericardiocentesis for pericardial effusion; surgical resection of RV aneurysm	Moderate pericardial effusion medically managed	None
RV function after stage I	Normal	Normal	Normal
Maximum interstage BNP (*pg*/mL)	894.5	2,072.4	457.7
Stage II operation	Glenn operation	Hemi-Fontan with PA plasty	Glenn operation
Stage II complications	None	None	Small pleural effusions
RV function after stage II	Mild/moderate decrease	Moderate decrease	Normal

serum brain natriuretic peptide (BNP) level obtained prior to discharge, he was started on oral digoxin. His function remained normal on serial evaluations during the interstage period. His stage II surgery (bidirectional Glenn) was uncomplicated with normal function at time of discharge; he was not continued on digoxin. He did require continued enteral diuretics for small pleural effusions upon discharge, which were weaned in the outpatient setting over the following weeks.

All three patients are now awaiting their Fontan completion operation (Table 1).

5 | DISCUSSION

LVNC, a genetic form of cardiomyopathy, has been well described in patients with and without associated congenital heart disease.[6,7] However, RVNC as an isolated disease has been reported in children only in isolated case reports.[8–10] Even fewer case reports have been published demonstrating RVNC in congenital heart disease, and has been limited to two-ventricle heart disease.[11] To our knowledge, this case series is the first to describe RVNC in HLHS. We have described patients with ventricular dysfunction (patients A and B) or "heart failure" as measured by elevated BNP (patient C) along their clinical course. All of the patients received additional medical therapies, such as digoxin or angiotensin converting enzyme-inhibitors (ACE-I, such as captopril or enalapril) to treat heart failure.[12]

Interestingly, two of our patients (A and B) had effusions (both pericardial and pleural) after surgery that required additional management. There is no data to suggest that pericardial effusions are associated with noncompaction, as most clinical reviews cite either absence of symptoms or heart failure symptoms as the primary clinical findings. Patient A required pericardiocentesis after stage I, and patient B required drainage of pleural and pericardial effusions after her stage II operation. These findings are unusual as pericardial effusion requiring intervention is a rare complication after the Norwood operation.[13] Patient A went on to have an even more complicated course with a very rare complication after pericardiocentesis: a ventricular pseudoaneurysm. Pseudoaneurysm of the RV infundibulum after RV-PA conduit placement, while rare, has been documented.[14] In contrast, the pseudoaneurysm in patient A was apical, and possibly due to intervention with pericardiocentesis and possibly because of the vulnerability of the myocardium because of the RVNC. There are rare case reports of pseudoaneurysm formation after pericardiocentesis.[15,16] Our patient required a repeat cardiac operation due to this complication with significant impact on the patient's clinical course.

Patients A and B also had recoarctation of the aorta found during routine prestage II catheterization. Known to be a cause of worse ventricular function in patients with single ventricle congenital heart disease, the role of coarctation in these patients with underlying abnormal myocardium is difficult to assess. With that said, after relief of coarctation with catheter-based intervention, these two patients continued to have some degree of RV dysfunction including after stage II operation, possibly related to their underlying abnormal myocardium. The longer term impact of abnormal myocardium remains to be seen as these patients await their Fontan completion and beyond.

From our perspective, patients with HLHS should be evaluated for possible RVNC with careful assessment of the myocardium during postnatal evaluation and postoperatively, especially in cases of abnormal RV function. Because criteria does not exist explicitly for RVNC, we continue to use the previously published recommendations for identification of NC in cases of the LV, with compaction to noncompaction ratios of more than 2:1. With enhanced knowledge of these patients at our institution, we will continue to identify these patients and gather additional criteria that could aid in the diagnosis of RVNC.

Based on our experience with these patients, RVNC in association with HLHS should be closely monitored for ventricular dysfunction and pericardial effusions after surgery, as there are significant clinical implications. Our understanding of this disease process is limited and requires additional investigation, but emphasizes the importance of appropriate diagnosis to allow for timely follow-up and counseling for this unique patient population.

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CONFLICT OF INTEREST

None.

AUTHORS CONTRIBUTIONS

Both authors participated in data analysis and approval of this article.

Concept/design, data collection and analysis, drafting: Gardner, Cohen

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