Use of pulsatile ventricular assist devices (VAD) is a well-established strategy for supporting children with end-stage heart failure (HF), as a bridge to heart transplant. In 2005, Stiller and colleagues1 described a cohort of 73 pediatric patients managed using Excor pumps (Berlin Heart Inc, Woodlands, Tex), of whom 12.5% were successfully weaned of support. Since then, there have been multiple reports of successful pulsatile VAD weans in children.2,3 Myocardial recovery has been usually reported within a window of 1 to 2 weeks, on extracorporeal support and longer on VAD support.3-5 Here, we describe 2 patients with HF requiring biventricular VAD (BiVAD) support, successfully weaned 2 and 6 months after VAD implantation.

CASE 1
A 6-month old girl (7 kg) was transferred for severe HF status postrepair of a late diagnosis of severe coarctation of the aorta (CoA) with minimal residual gradient and requiring escalating inotropic support. Echocardiography demonstrated severely diminished biventricular systolic function and left ventricular end diastolic volume (LVEDVi) of 87.14 mL (z score, 8.02). The etiology for HF was likely multifactorial: late repair of severe CoA, ischemic insult postbypass. Due to severe biventricular dysfunction, BiVAD support was initiated using Excor 10 mL pumps based on our institutional approach. LV biopsy at implant demonstrated interstitial fibrosis with no inflammation, patchy cardiomyocyte atrophy, and hypertrophy of LV trabeculae (Figure 1, A). Surveillance echocardiograms were performed. Normalization of right ventricular function was noticed at 2.5 months. A gradual decrease in LVEDVi was noted over the next 2 months to z score of +2 with improvement in LV function to normal at 4.5 months. Biventricular function was monitored over 4 weeks, with no evidence of deterioration. A weaning trial was performed (Table 1) and good hemodynamic parameters were measured by...
cardiac catheterization (Table 2). VAD was explanted with good clinical function (Table 3). Patient was changed to Status 7 (inactive) once evidence of recovery was seen. This patient is currently 21 months postexplant with normal function and no recurrent CoA (LVEDVi after explant was 23.44 mL).

### TABLE 1. University of Florida ventricular assist device (VAD) weaning protocol (heparin bolus 75 IU/kg, 15 minutes before VAD action)

| Day of wean | VAD action | Parameters monitored |
|-------------|------------|----------------------|
| 1           | BiVAD rate decreased by 50% for 30 min | Vital signs (heart rate, respiratory rate, blood pressure) every 3 min, Continuous NIRS, Mental status, SvO₂ measurement every 10 min, Echocardiogram every 10 min (left ventricular end diastolic volume, ejection fraction, and qualitative assessment of wall function) |
| 2           | BiVAD rate decreased by 75% or 35 (whichever was higher) for 30 min; following this, BiVAD completely paused for 3 min | Important: Both pumps have to be manually pumped to fill and eject every 30 s, Same as Day 1, SvO₂ measurement every 10 min and at the end of pause, Echocardiogram every 10 min and at the end of the pause (size and function) |
| 3           | BiVAD rate decreased by 75% or 35 (whichever was higher) for 30 min; following this, BiVAD completely paused for 6 min | Important: Both pumps have to be manually pumped to fill and eject every 30 s, Same as Day 2 |
| 4           | BiVAD rate decreased by 75% or 35 (whichever was higher) for 30 min; following this, BiVAD completely paused for 10 min | Important: Both pumps have to be manually pumped to fill and eject every 30 s, Same as Day 2 |
| 5- Right heart catheterization | BiVAD rate decreased by 75% or 35 (whichever was higher) for 30 min; following this, BiVAD completely paused for 10 min | Important: Both pumps have to be manually pumped to fill and eject every 30 s, Same as Day 2 with hemodynamic measurements in catheterization include right atrial, right ventricular end diastolic pressures, and pulmonary venous wedge pressures in addition to estimating cardiac output |

VAD, Ventricular assist device; BiVAD, biventricular assist device; NIRS, near-infrared spectroscopy; SvO₂, mixed venous oxygen saturation.
CASE 2

A 9-month-old girl (8 kg) was admitted with cardiogenic shock (biventricular dysfunction due to dilated cardiomyopathy), with initial LVEDVi of 87.2 mL (z score, 7.79).

BiVAD support was initiated using Excor 10 mL pumps as a bridge to heart transplant. A biopsy from LV at VAD implantation showed cardiomyocyte size variation with patchy interstitial fibrosis without inflammation. (Figure 1, B). On surveillance, recovery in LV function with decrease in LVEDi was noticed at 4 weeks with recovery of right ventricle. LV function was normal at 6 weeks and was monitored for another 2 weeks. Weaning trial was performed at 8 weeks and was tolerated. The patient underwent successful a VAD explant. Follow-up echocardiogram showed LVEDVi of 24.51 mL (z score, 0.7) and currently the patient has normal biventricular function 18 months postexplant (Table 3).

DISCUSSION

Although recovery is well described in the adult population on VAD support, there is limited data in children using pulsatile VADs, with some of the case reports of recovery being on continuous flow devices.3-8 In 2016, Hezter and colleagues3,9 described that 15% (n = 18) of patients had recovery over a median time frame of 1 to 2 months. Ihnat and colleagues5 reported a higher frequency of recovery than other cohorts—61% (8 out of 13)—with the longest duration of support before wean being 21 days. Recovery was seen more frequently in myocarditis and idiopathic HF, with mean patient age <2 years.5

Current evidence suggests initial myocardial injury could arise from high levels of transforming growth factor beta and imbalance of the renin-angiotensin system, which promotes interstitial fibrosis and LV dilation with myocyte hypertrophy, respectively.10 Reverse modeling with VAD support has been seen in adults over longer time periods, leading to reduction in myocyte size and the area of fibrosis.11

Our imaging surveillance protocol includes echocardiogram twice a month for the first 3 months and monthly thereafter, monitoring LV dimensions, ejection fraction, and wall motion assessment. If wall motion recovery is noted, then imaging is performed weekly. Once function is identified as low normal qualitatively, ejection fraction >45% to 55% with LVEDVi within +2 z scores, a VAD wean trial is initiated after ensuring recovery is sustained over the next 2 to 4 weeks. Recurrence of HF postexplant has been described by Irving in 3 of 11 patients, requiring VAD reimplant and heart transplant. The etiology in all 3 was dilated cardiomyopathy.5 We suggest an active surveillance protocol for any signs of myocardial recovery in all pediatric patients with a VAD.

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| Patient | LVEF (at diagnosis) | RV function at diagnosis | LVEDi (cm) at diagnosis | LVEF at VAD wean | RV function at VAD wean | LVEDi (cm) at VAD wean |
|---------|---------------------|--------------------------|-------------------------|------------------|------------------------|------------------------|
| 1       | 8%-10%              | Severely depressed       | 8.4 (z score, +8)       | 55%-60%          | Normal                 | 3 (z score, +1.5)      |
| 2       | 5%-7%               | Mildly depressed         | 4.4 (z score, +9)       | 60%-65%          | Normal                 | 2.34 (z score, –1)     |

LV, Left ventricle; EF, ejection fraction; RV, right ventricle; LVEDi, left ventricular end diastolic dimension; VAD, ventricular assist device.
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