Images in Cardiovascular Medicine

Extracorporeal Membrane Oxygenation Support in Severe Hypertrophic Obstructive Cardiomyopathy Associated With Persistent Pulmonary Hypertension in an Infant of a Diabetic Mother

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A male infant was delivered at 37 weeks of gestation via cesarean delivery because of minimal fetal heart tone variability and late fetal decelerations to a 31-year-old mother whose pregnancy was complicated by maternal type 2 diabetes mellitus and chronic hypertension. Fetal echocardiography had been performed at 20 weeks of gestation, which showed a structurally normal heart without ventricular hypertrophy (Figure 1 and Movie IA and IB in the online-only Data Supplement). The mother had been treated previously with insulin, and during pregnancy, had hemoglobin A1c of 7.9% (diabetic patients with adequate glycemic control have values <7% to 8%).1 The birth weight was 4.8 kilograms. The infant developed respiratory failure within the first 15 minutes of life, requiring intubation and mechanical ventilation. The umbilical cord venous blood gas demonstrated a pH of 7.16 and partial pressure of oxygen of 31 mm Hg. A transthoracic echocardiogram (TTE) was performed following the patient’s admission to the neonatal intensive-care unit because of auscultation of a grade-IV/VI harsh systolic ejection murmur heard throughout the precordium. TTE showed severe biventricular hypertrophy, mild right ventricular outflow tract obstruction, severe left ventricular outflow tract obstruction with a peak velocity of 5.0 meters per second, almost complete obliteration of the left ventricular outflow tract, a patent foramen ovale with left-to-right shunting, a large patent ductus arteriosus (PDA) with bidirectional shunting, and rightward septal bowing throughout systole (Figure 2 and Movie IIA–IIC in the online-only Data Supplement). These findings were consistent with severe hypertrophic obstructive cardiomyopathy associated with infants of diabetic mothers (HOCM-IDM) and concomitant persistent pulmonary hypertension of the newborn (PPHN). HOCM-IDM is a well-recognized cardiomyopathy found in fetuses and infants at birth and is attributable to hyperinsulinism as a result of maternal gestational diabetes. It is characterized on echocardiogram by ventricular hypertrophy with greater septal hypertrophy than free-wall hypertrophy as well as resultant biventricular outflow tract obstruction.2 It is classified as a hypertrophic cardiomyopathy and distinguished from other forms of hypertrophic cardiomyopathy by its transient and nonfamilial nature in the setting of hyperinsulinism.3

The patient progressed with severe hypoxemia, requiring high conventional ventilatory support as well as circulatory failure indicated by hypotension responsive only to substantial fluid administration to maintain preload (requirement of >1.2 L of fluid in the first 24 hours of life). Medical management also included intravenous β-blockade for the dynamic outflow tract obstruction and inhaled nitric oxide for the PPHN. As a result of refractory severe hypoxemia and hypotension as well as development of oliguria and lactic acidosis, the decision was made to proceed with Extracorporeal Membrane Oxygenation (ECMO) cannulation on day 4 of life. After 6 days of support, ECMO was weaned, with the rationale that sufficient time was given for the PPHN to have improved. The PDA closed spontaneously during the period of ECMO support. Members of the care team and family agreed that further lifesaving measures would not be offered in the event of cardiac failure resulting from ECMO wean. After the procedure, clinical improvement was evident with resolution of pulmonary hypertension and lessened degree of left ventricular outflow tract obstruction, although significant biventricular hypertrophy was still evident on TTE. The patient was discharged from the hospital at 2 months of life on oral β-blockade. Repeat TTE at 3 months of age showed significant improvement in septal hypertrophy, biventricular outflow tract obstruction, and septal configuration (Figure 3 and Movie III A–IIIC in the online-only Data Supplement).

The natural history of HOCM-IDM appears to be benign, with resolution of septal hypertrophy within 2 to 12 months.2 ECMO support has been used in this disease, although there are few data on its effectiveness and the appropriateness of its use in a condition that regresses over a period of months.4 This case demonstrates a severe example of the disease, which

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The online-only Data Supplement is available with this article at http://circ.ahajournals.org/lookup/suppl/doi:10.1161/CIRCULATIONAHA.114.010678/-/DC1.

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(Circulation. 2014;130:1923-1925.)

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Circulation is available at http://circ.ahajournals.org

DOI: 10.1161/CIRCULATIONAHA.114.010678
developed after a normal fetal echocardiogram at 20 weeks of gestation, further complicated by PPHN. ECMO support allowed for time to ameliorate the pulmonary hypertension and support the patient during this critical period, with subsequent improvement of the ventricular hypertrophy, as expected in the natural history of HOCM-IDM.

Disclosures

None.

References


Figure 3. Transthoracic echocardiogram (TTE) at 3 months of life. A, Parasternal long-axis view showing interval improvement in left ventricular outflow tract patency. B, Parasternal long-axis view with color-flow Doppler showing improvement of flow through the left ventricular outflow tract and of mitral regurgitation. C, Parasternal short-axis view showing improvement of left ventricular hypertrophy. D, Continuous-wave Doppler through the left ventricular outflow tract showing a velocity of 2.9 meters per second (improved from 5.0 meters per second). Ao indicates aorta; IVS, interventricular septum; LA, left atrium; LVOT, left ventricular outflow tract; MR, mitral regurgitation; MV, mitral valve; and PW, posterior wall.
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Circulation. 2014;130:1923-1925
doi: 10.1161/CIRCULATIONAHA.114.010678
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/130/21/1923

Data Supplement (unedited) at:
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