Severe cardiac failure in newborns with VGAM

Prognosis significance of hemodynamic parameters in neonates presenting with severe heart failure owing to vein of Galen arteriovenous malformation

Abstract

Objective: Neonatal vein of Galen malformation complicated by severe cardiac failure is a rare disease. The purpose was to assess the outcome of this life-threatening malformation and identify hemodynamic prognostic factors.

Design: Retrospective study.

Patients: Twenty-four newborns with cardiac failure requiring mechanical ventilation were consecutively admitted from 1986 to 2000.

Interventions: Cardiovascular evaluation including echocardiogram was performed in all cases. Eighteen transarterial shunt occlusions with glue were applied by the same team of three physicians.

Measurements and results: Twelve babies survived and underwent one endovascular session at least (median age 20 days) with a mean 63 months follow-up. Embolization was not performed in 6 of the 12 nonsurvivors because of severe brain damage or profound hypotension Cardiogenic shock occurred in all nonsurvivors, but also in one long-term survivor (p=0.01). Echocardiogram showed signs of right ventricular failure, most often in the dead babies (p=0.005). The pulmonary systemic arterial pressure ratio was significantly higher in the nonsurvivor group (p=0.031), and it decreased significantly after the first embolization only in patients who survived (p=0.01). Patent ductus arteriosus and a diastolic aortic reversed-flow were present in all nonsurvivors in contrast to 30% of the long-term survivors (p=0.003). There was no difference in the left ventricular contractility and mean cardiac output between the two groups.

Conclusions: The outcome of vein of Galen malformation complicated by severe cardiac failure requiring mechanical ventilation remains poor. Neonatal embolization seems to be beneficial only in babies without suprasystemic pulmonary hypertension.

Keywords Newborn · Severe cardiac failure · Vein of Galen malformation · Persistent pulmonary hypertension of the newborn · Encephalomalacia · Ductus arteriosus · Embolization

Introduction

Vein of Galen aneurysmal malformation (VGAM) is a rare but potentially life-threatening cerebral vascular malformation. It was the first cerebrovascular malformation recognized to be of embryonic origin. It develops from the median vein of the prosencephalon [1]. Symptoms may vary, depending on the patient’s age. There is congestive heart failure and encephalomalacia in neonates, macrocrania and hydrocephalus in infants and minimal cardiac insufficiency, asymptomatic cardiac enlargement, headaches, or intracranial hemorrhage in older children. Neurodevelopmental delay or convulsions may occur at any age, but with different mechanisms and pathophysiology [1].
First described by Gold et al., VGAM revealed by severe cardiac failure remains a rare neonatal disease, often having a fatal outcome or complicated by severe brain damage [2, 3]. Since the introduction of transarterial endovascular occlusive procedure with glue, the survival rate and incidence of neurologic complications have significantly improved over the last 15 years [1, 4]. The improvement in fetal and postnatal ultrasound techniques allows an earlier diagnosis of VGAM and prompt management of these patients. This has encouraged neonatologists to refer neonates with severe cardiac failure to an interventional neuroradiology team earlier than in the past [5]. However, there is still controversy as to the ideal timing of endovascular management. In this study, we assessed the outcome and evaluated the hemodynamic profile of neonates with VGAM complicated by severe cardiac failure in order to identify possible prognostic factors.

Material and methods

From October 1986 to June 2000, 258 patients with VGAM were hospitalized at our institution for evaluation and treatment. Data were analyzed retrospectively, using the patient’s chart, following a database system. The analysis was limited to a group of newborns with proven VGAM, who were admitted to the pediatric intensive care unit (PICU) with severe cardiac failure requiring mechanical ventilation. Severe cardiac failure was defined clinically by the presence of tachycardia, respiratory distress, and hepatomegaly. Cardiogenic shock was defined by the presence of clinically poor peripheral perfusion, oliguria and the need for inotropic support, independently of the level of systemic arterial blood pressure or biochemical markers of organ failure. The neurologic evaluation included a physical examination, EEG, CT scan and MRI. Laboratory tests included arterial blood gases, PT and PTT, CBC with platelet count, plasma lactate level, and liver and renal function tests to assess the presence of peripheral organ injury (data not shown).

A cardiac ultrasound evaluation was performed in every patient at the time of admission and repeated a few days after each endovascular procedure. It included the measurement of the right and left end-diastolic diameters (RVEDD, LVEDD), left ventricular shortening fraction (LVSF), stroke volume (SV), left ventricular output (reflecting cardiac output or CO as the product of velocity time integral by the cross-sectional area at the aortic valve level and multiplied by the heart rate), and systolic pulmonary arterial pressure, based on measurement of the tricuspid flow regurgitation with the modified Bernouilli equation. Cardiac malformations were ruled out as well as the presence of a patent ductus arteriosus (PDA) and a patent foramen ovale (PFO). The ductal flow pattern were analyzed according to pulsed Doppler sampling placed in the color area of the highest velocity. Descending aortic reverse flow, aortic flow reversal, and/or evidence of severe brain damage were analyzed according to pulsed Doppler sampling placed in the color area of the highest velocity. Descending aortic flow reversal was researched at the level of the PDA insertion. The shape of the interventricular septum was described, based on the right-to-left side motion, as: 1=normal, 2=intermediate, and 3=complete right-to-left shift with LV collapse.

The presence of brain damage or encephalomalacia was considered to be a contraindication for the endovascular procedure, and the patient’s eligibility for this procedure was based upon our previous results [1, 6, 7]. Conversely, when cardiac function improved following the initial management, the embolization procedure was postponed until the baby was 5 months old. The endovascular procedure consisted in catheterization of the femoral artery and a first transarterial occlusion session using N-butyl cyanoacrylate. The goal for this first session was to reduce the shunt by a third, based on the size of the aneurysm [8].

Patients were divided into two groups based on outcome: survivors and non-survivors. Fisher’s exact test and Wilcoxon rank-sum test were used for statistical analysis. A p<0.05 was considered significant.

Results

Ninety-six newborns diagnosed with VGAM were hospitalized at the Bicêtre Hospital between October 1986 and June 2000. Twenty-four of them (25%), including 19 who were hospitalized after 1992, were admitted to the PICU because of cardiac failure and the need for mechanical ventilation. Fifteen were male, and the median postconceptional age was 40 weeks. Twelve patients died during hospitalization (including 1 before 1992) (non-survivor group, NS). Six of them were not embolized because of impossible technical challenges (severe cardiogenic shock with hypotension and aortic reverse flow, n=3) or evidence of severe brain damage (n=3) and died. Six more patients died after the first embolization because of multiple organ failure and intractable cardiac failure. Twelve patients (50%) underwent a first successful embolization of the VGAM and were then discharged (survivor group, S). There was no difference between the S and NS groups with respect to postconceptional age, sex, birth weight, and head circumference (Table 1). The procedure was done at 21 (7–38) days of life with no difference between the S and NS groups (26 vs 20 days, respectively, p=ns). The diagnosis of VGAM was made in utero by fetal ultrasound in three babies by the third trimester of pregnancy. Two of them also had a fetal MRI. The median age at the time of VGAM diagnosis in the remaining 21 babies was 3 days (0–15) with no differences between the S and NS group (Table 1). There was also no difference with respect to the time of appearance of clinical signs of cardiac failure and the beginning of mechanical ventilation between the two groups (Table 1).

All babies presented clinical signs of severe cardiac failure at the time of admission, with tachycardia, tachypnea, and hepatomegaly. Most of them had a cervical “dancing” carotid pulse and distended jugular veins. Every patient presented a peripheral pulse and precordium bounding owing to a hyperdynamic status with increased venous return and right heart overload at the echocardiogram (Table 2). The initial treatment consisted of continuous enteral feeding, a diuretic (furosemide 2–6 mg/kg per day) to reduce the preload and mechanical ventilation. Every patient received opioids and benzodiazepines to lower oxygen consumption. The hemodynamic management was then based on the echocardiogram findings. An infusion of dobutamine was started in 13 patients because of normal- to low-cardiac output, poor peripheral perfusion, and/or low urinary output and severe right cardiac failure. Seven patients received additional dopa-