Aortic Diastolic Pressure Decay in Congenital Arteriovenous Malformations

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SUMMARY. We describe three infants with large congenital arteriovenous malformations, two intracranial and one intrathoracic, who have high output congestive heart failure but normal pulse pressures. The theoretical basis of these pulse pressure findings in contrast to those in infants with shunting secondary to the patent ductus arteriosus is presented.

KEY WORDS: Congenital arteriovenous malformations — Aortic diastolic pressure decay — Patent ductus arteriosus — Neonate

Case Reports

Case 1

This 3400-g female infant born following a full-term gestation had Apgar scores of 2 and 2 at 1 and 5 min, respectively. Resuscitation included intubation and assisted ventilation. Following stabilization, increased precordial activity, a heart murmur, and hepatomegaly were noted. A faint carotid bruit was audible but none was detected over the head or abdomen. The blood pressure was 52/25 in the upper and lower extremities. The chest radiograph revealed bilateral pleural effusions, as well as indistinct cardiac borders. In spite of digitalization and diuretic therapy, the infant remained in congestive heart failure at 48 h. Cardiac catheterization revealed increased oxygen saturation (87%) in the superior vena cava with no evidence of intracardiac shunting by oximetry, pulmonary artery hypertension (61/28 mmHg), and a complex arteriovenous malformation at the base of the skull along with an aneurysmal sac filling most of the posterior fossa with very little blood flow to the cerebral hemispheres. The aortic blood pressure at catheterization was 62/40 and the aortic diastolic half-time determined using a previously described method [7] was 800 ms (increased). Other catheterization results included: increased PVR (Res units) = 18.1; normal SVR (Res units) = 22.5; increased C.O. (L/min) = 1.8; increased Cardiac Index (L/min/m²) = 7.8; and normal O₂ consumption (cc/min) = 35.

This infant was subsequently treated with two embolization procedures with only moderate improvement in her congestive heart failure. Complications included embolization to the lung resulting in pulmonary necrosis and pneumothoraces and subsequent pseudomonas sepsis. Although the intracranial flow decreased markedly, congestive heart failure persisted. The infant expired at 5 weeks of age.

Case 2

This 2260-g female infant born following a full-term gestation had Apgar scores of 7 and 7 at 1 and 5 min, respectively. No resuscitation was required; however, persistent cyanosis and respiratory distress necessitated transfer to the neonatal intensive care unit. Precordial activity and pulses were markedly increased. A grade 4-5/6 continuous murmur was present across the upper chest with diffuse radiation. In addition, an S3 gallop was audible. The blood pressure was 60/40 mmHg. The chest radiograph revealed diffuse cardiomegaly with normal to decreased pulmonary vascularity. Cardiac catheterization revealed insignificant oxygen saturation differences (77-81%) in the superior vena cava, right atrium, right ventricle, left atrium, and left ventricle. Marked tricuspid insufficiency and a large right-to-left shunt across a patent foramen ovale were present. These findings were supportive of the diagnosis of persistent pulmonary hypertension of the newborn and could account for the multichamber desatu-
ratriation. At catheterization, the aortic pressure was 75/45 and the aortic diastolic half-time was 500 ms (normal) [7]. An angiogram of the left ventricle revealed a dilated ascending aorta, dilated left subclavian artery, little contrast distal to the left subclavian artery, and early recirculation of contrast into the superior vena cava and right heart. A large arteriovenous malformation from the left subclavian artery to the left subclavian vein was identified on angiography. The lesion was treated with ligation of the infant’s left subclavian artery and catheter-coil embolization of some residual arteriovenous malformations. Aside from a shortened left arm and no significant palpable pulse to the left arm at 6 years, the patient has been asymptomatic from a cardiovascular standpoint.

Case 3

This 3400-g female infant born following a full-term gestation had Apgar scores of 7 and 9 at 1 and 5 min, respectively. No resuscitation was required. By 24 h of age, the infant had progressive respiratory distress. A murmur was audible over the left sternal border and radiated to the apex, axilla, and neck with an audible bruit over the head, particularly the anterior fontanelle. The chest radiograph revealed diffuse cardiomegaly. The blood pressure was 70/45. Aortic diastolic half-time was 477 ms (normal). An arteriogram, performed at 5 days of age, revealed an arteriovenous malformation draining from posterior choroidal, lenticulostriate, and pericallosal arteries into an enlarged vein of Galen and straight sinus. The infant was digitalized but subsequently developed progressive congestive heart failure. At 4 months of age, the infant developed hydrocephaly requiring shunting. At that time, some vascular feeders to the malformation were clipped. At 7 months of age, progressive ventriculomegaly recurred requiring shunt revision. Additional clipping of feeders to the malformation was considered; however, progressive deterioration ensued over the subsequent month. No further intervention for the malformation was considered appropriate and the infant expired by 8 months.

Discussion

Patients with chronic arteriovenous malformations often have high-output cardiac failure [4]. The failure may not manifest acutely but develops as compensatory increases in blood volume occur to counteract the lower blood pressure and shift of blood from the normal circulation into the malformation [4]. Cumming described this phenomenon in two neonates with cerebral arteriovenous malformations who presented in congestive heart failure and were found to have cardiac outputs greater than twice normal [2]. Similarly, in Case 1 of this report, catheterization data demonstrated a significant increase in cardiac output, stroke volume, and presumably blood volume since the heart rate was in the normal range. We do not have hemodynamic data regarding cardiac outputs in Cases 2 and 3 but would have expected similar results in view of their diffuse cardiomegaly and normal blood pressures. Theoretically, these “normal” blood pressures may actually represent elevated pressures in the presence of arteriovenous malformations and may represent increased systemic vascular resistance in response to the cardiac failure due to volume overload [8].

Other neonates with cerebral arteriovenous malformations and congestive failure have been shown to have myocardial ischemia or infarction [5]. Both volume and pressure overload may be contributing to the failure [5]. In our series, autopsy findings in Cases 1 and 3 demonstrated ventricular hypertrophy without infarction.

The increase in blood pressure, particularly the diastolic component, in our patients with arteriovenous malformations is in distinct contrast to the acute changes noted in the premature neonate with left-to-right shunting across the patent ductus arteriosus where widened pulse pressures with low diastolic pressures predominate [7]. In our three cases, widened pulse pressures and rapid aortic diastolic pressure decays were clearly absent. Similarly, the two cases reported by Cumming had relatively narrow pulse pressures (Patient 1 had a BP of 61/40 and Patient 2 had a BP of 77/50) [2] and were thus unlikely to have rapid aortic diastolic pressure decay.

What is the basis for the difference in pulse pressure and rate of aortic diastolic pressure decay in the infant with a patent ductus arteriosus and an arteriovenous malformation? When no shunts are present, aortic diastolic pressure decays exponentially according to the first-order equation: $P = P_o e^{-\frac{t}{\tau}}$, where $P$ represents the diastolic pressure at time, $t$, $P_o$ represents the initial diastolic pressure, and $\tau$ represents the aortic diastolic pressure time constant [1, 6, 7]. The aortic diastolic pressure time constant, $\tau$, can be approximated by the product of resistance, $R$, and capacitance, $C$, of the arterial system. With the exception of the aorta (with the primary function of conductance) and the capillaries (with the primary function of exchange) vascular segments throughout the systemic and pulmonary circulations have both resistive and capacitive functions [3]. The relative resistive and capacitive functions in each segment are not necessarily of equal importance or magnitude but are determined by the structural differences [3]. In both the systemic and pulmonary circulations, the primary function of the arteries and arterioles is resistive, the small veins and venules is capacitive, and the large veins and vena cava is resistive [3]. In the absence of any shunts (see Fig. 1), the proximal arterial and arteriolar resistive properties are the main determinants of $\tau$. The capacitive venous compartment is too far downstream to affect $\tau$. 