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Fludrocortisone in the Management of an Infant with a Single Ventricle and an Overly Tightened Pulmonary Artery Band

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Abstract We describe a 2-month-old female infant with a univentricular heart and overly tightened pulmonary artery band (PAB) resulting in hypercyanotic episodes. Despite adequate doses of morphine and a beta-blocker, she continued to have multiple hypercyanotic events per day. Treatment with fludrocortisone resulted in resolution of these episodes and allowed performance of a Glenn shunt at 3 half months of age. We conclude that fludrocortisone may be beneficial in the management of an overly tightened PAB until the patient is suitable for surgical intervention.

Keywords Fludrocortisone · Pulmonary artery band · Congenital heart disease

Introduction

Pulmonary artery banding (PAB) is an important surgical procedure in the palliation of congenital heart disease. It is used as a preparatory step for cavopulmonary anastomosis (CPA) to ensure normal pulmonary artery pressure and resistance, particularly in patients with univentricular heart pathophysiology [2]. The management of an overly tightened PAB is difficult and may result in forcing earlier than optimal surgical intervention toward CPA with possible detrimental outcome [4].

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Fludrocortisone is a corticosteroid with potent mineralcorticoid and minimal glucocorticoid activity. It has been used in the treatment of primary adrenocortical insufficiency and salt-losing adrenogenital syndromes and is effective in the treatment of idiopathic orthostatic hypotension in combination with salt and volume supplements [13]. We report the use of fludrocortisone in the successful management of a patient with a univentricular heart and overly tightened PAB. Its use allowed a period of growth until the patient was suitable for CPA.

Case Report

Using fetal echocardiography, we diagnosed a female infant with congenital heart disease at 21 weeks of gestation. This 3.68-kg infant was born at term, and her postnatal echocardiogram confirmed her fetal cardiology evaluation as coarctation of the aorta, univentricular heart (double-inlet right ventricle with severe mitral stenosis), double-outlet right ventricle, patent ductus arteriosus, and patent foreman ovale. The decision was made to delay a bidirectional Glenn procedure until 3 months of age, as practiced at our institution and shown by others to be the earliest, safest age for this procedure [4]. Her initial management included prostaglandin E_1 until she underwent surgical repair of the coarctation at 7 days of age (weight, 3.50 kg) by extended end-to-end anastomosis and PAB. During the next 3 weeks, echocardiogram indicated that the patent foreman ovale became increasingly restrictive and the PAB migrated to the left pulmonary artery, thus preferentially shunting blood to the right pulmonary artery. At a cardiac catheteriza-

tion done on day of life 20, the echocardiographic findings were confirmed and blade septostomy was performed. After this procedure, she developed a pericardial effusion, and 2 days later she had a surgical revision of her PAB, adjusted according to intraoperative Doppler velocity of 3.8 m/sec, and evacuation of a small hemopericardium. She then became stable and was managed with digoxin and furosemide until discharged from the hospital at 1 month of age (weight, 3.53 kg).

She returned to the emergency room 2 weeks later with vomiting and diarrhea and subsequently was admitted for management of dehydration (admit weight, 3.53 kg). After we stopped furosemide and optimized her hydration with aggressive intravenous fluids, she returned to baseline with oxygen saturations 65–80% on room air. On the third hospital day, she began having prolonged desaturation spells associated with listlessness and pallor. These episodes occurred two or three times per day with oxygen saturations by pulse oximetry ranging between 30 and 40%, despite 6 L/min of oxygen by mask. An echocardiogram showed findings consistent with an overly tightened PAB (a d/D ratio equal to 0.2 [2], which in our experience was the smallest for her weight and age category, and a maximal Doppler-derived velocity of 5 m/sec [27]), which was thought to be the cause of her hypercyanotic spells. She responded acutely to knee–chest positioning but continued to have one or two spells per day during the next 9 days, despite the use of morphine sulfate and increasing doses of propranolol up to 6 mg/kg/day, orally, divided every 6 hours.

After discussing the potential risks and benefits of using fludrocortisone with pediatric endocrinologists and subsequently with the parents, she was started on 0.05 mg once daily at 2 months of age (weight, 4.33 kg). During the next 4 days, she had only two hypercyanotic episodes requiring knee–chest positioning and her average oxygen saturation remained in the mid-70 s. Her fludrocortisone dose was then increased to 0.1 mg once daily, after which she stopped having any hypercyanotic spells, maintained oxygen saturations > 80%, and weaned off oxygen support (Table 1). She was discharged home 9 days later (weight, 4.62 kg) on fludrocortisone and propranolol at the dosages previously.

At the age of 3 and half months, she underwent cardiac catheterization (admit weight, 5.30 kg), which confirmed the diagnosis of overly tightened PAB and partially restrictive patent foreman ovale with the presence of bilateral subarterial coni and no residual coarctation. An echocardiogram done at that time showed a wide-open subpulmonic area, verifying that a stenosis at this level was not contributing to her hypercyanotic spells. During the same admission, she underwent surgical creation of Damus–Kaye–Stansel connection, atrial septectomy, and placement of a Glenn anastomosis. At the age of 1 year, she was doing very well with normal neurological status. There was no evidence of complications from the 1 and half months of fludrocortisone.

Table 1 Comparison of the lowest recorded saturation and mean oxygen requirement averaged over the 8 days before and 8 days after treatment with fludrocortisone

	Lowest saturation (%)	Oxygen requirement (L/min)
Before	52.25	2.97
After	62.75*	2.06**

*$p < 0.02$.

**$p < 0.05$.

Discussion

PAB is a palliative surgical technique used in a staged correction of congenital heart defects. This technique is used in children born with cardiac defects characterized by pulmonary overcirculation and elevated pulmonary pressure who cannot undergo surgical repair or palliation in infancy. The prime example is a patient with univentricular heart. The main objective of PAB is to reduce excessive pulmonary blood flow and ensure normal pulmonary pressure and resistance.

One of the most difficult tasks for the congenital heart surgeon when placing a PAB is to balance its ''tightness.'' In a small infant in whom an overly tightened PAB has been placed, the potential treatment is either surgical readjustment of the PAB or medical therapy. Surgically readjusting the PAB may carry the same or greater risk of PAB. Risks include creation of dysplastic changes in the pulmonary valve leaflets, obstruction of coronary blood flow, erosion of the band into the pulmonary artery, ineffectual restriction of the pulmonary blood flow, and impingement or stenosis of one or both of the branch pulmonary arteries. The medical treatment of an overly tightened PAB is difficult and is directed toward increasing the systemic vascular resistance and afterload to reverse or decrease the intracardiac right-to-left shunting of blood.

Fludrocortisone is a salt-retaining steroid with a very low profile of side effects. It has two major mechanisms by which it increases systemic afterload. The first is retention of sodium, causing an expansion of plasma

volume. The second is sensitization of α-adrenergic receptors, thereby increasing the sympathetic response to norepinepliine. These effects are transient and are limited to the period of its use. Because of these effects, fludrocortisone has been used successfully in the treatment of neurocardiogenic syncope [6]. The drug is usually tolerated well with rare side effects [1].

The salutary effect of fludrocortisone in raising the systemic vascular resistance was successfully used in the difficult medical management of our patient with an overly tightened PAB. The concept of increasing systemic vascular resistance to overcome pulmonary or subpulmonary obstruction in the setting of a univentricular heart, such as in hypercyanotic tetralogy of Fallot spells, is well addressed in the literature [5]. This increase in systemic vascular resistance was associated with a minimal overall increase in our patient's systolic blood pressure (with averaged daily ranges of 72.5–94.3 mmHg before and 73.3–98.9 mmHg after). Its use resulted in a prompt decrease in the severity and frequency of the hypercyanotic episodes, and by adjusting the dose these episodes were completely prevented. The use of this medication delayed the need for further surgical intervention, allowing our patient a period of growth until she was suitable for CPA.

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