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RESEARCH ARTICLE
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ANASTHETIC MANAGEMENT OF CIRCUMCISION IN A 4-YEAR - OLD CHILD WITH PULMONARY STENOSIS - A CASE REPORT

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INTRODUCTION

The largest category of human birth defects accounts for almost one percent of malformations among live birth abnormalities, including heart and vascular anomalies. 33 percent of children with chromosomal abnormalities have a congenital heart defect. Isolated pulmonary stenosis with an intact ventricular septum consists of 10 % of congenital heart defects, with further subdivision of valvular, supravalvular, and infravalvular lesions.

Of these valves, pulmonary stenosis is the most common, about 80-90% of the reported cases. This case report discusses a 4-year-old child with tight phimosis and straining for micturition, posted for circumcision, on echocardiogram, follow-up of pulmonary stenosis was detected at birth.

Case Report

The child was born as a full-term baby by C-section to nonconsanguineous parents with no antenatal complications in the mother. Routine echocardiogram revealed a dome-shaped pulmonary valve with no other morphological anomalies to fit into a syndrome. Milestones were normal. There was no breath holding or cyanotic spells. The child was advised for periodic echocardiographic surveillance for pulmonary pressure gradient and flow across the valve by Doppler. The interventricular and interatrial septae were intact with normal chamber configuration.

The child was asymptomatic. At the age of 1 year Doppler report showed PVG, suggesting mild to moderate {3.2/40mmHg valvar pulmonary stenosis with thick valve and restricted opening.

Clinical presentation at the age of three years, the child had normal mental state, no cardiorespiratory symptoms, no murmurs on auscultation, normal heart sounds, thickened pulmonary valve by echocardiogram and doppler showed PV max/PV max PG 2.53/25.64 mmHg, the child was 15 kgs, tight phimosis and otherwise normal.

Clinical Features	**Echo/Doppler {figure 1, 2}**		
Wt: 15 kgs, HR: 120/min		1 year	3 years
BP: 86/40mmHg, RS: clear	Chambers	normal	Normal
CVS: S1S2 present, Per abdomen: soft	IVS	Intact	Intact
HC: Normal, Eyes: normal,	PV	Thick	Thick
Limbs: normal,	PV V max	3.2	2.53
Hb: 11gm	PV max PG	40	25.6
Total counts: normal	EF	68%	67%

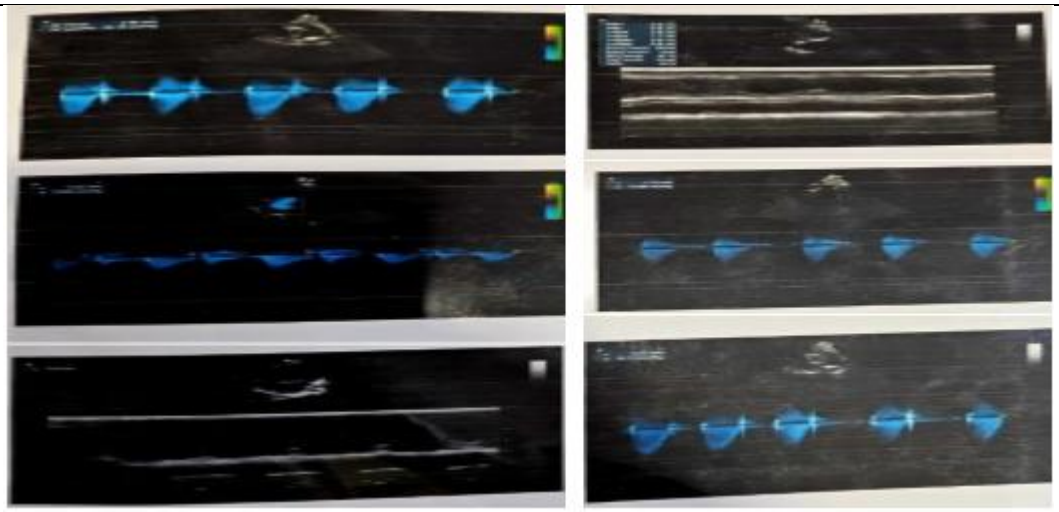

Figure 1: Doppler image across the pulmonary valve showing pulmonic valve PG 25.64 mmHg at 3 years of age

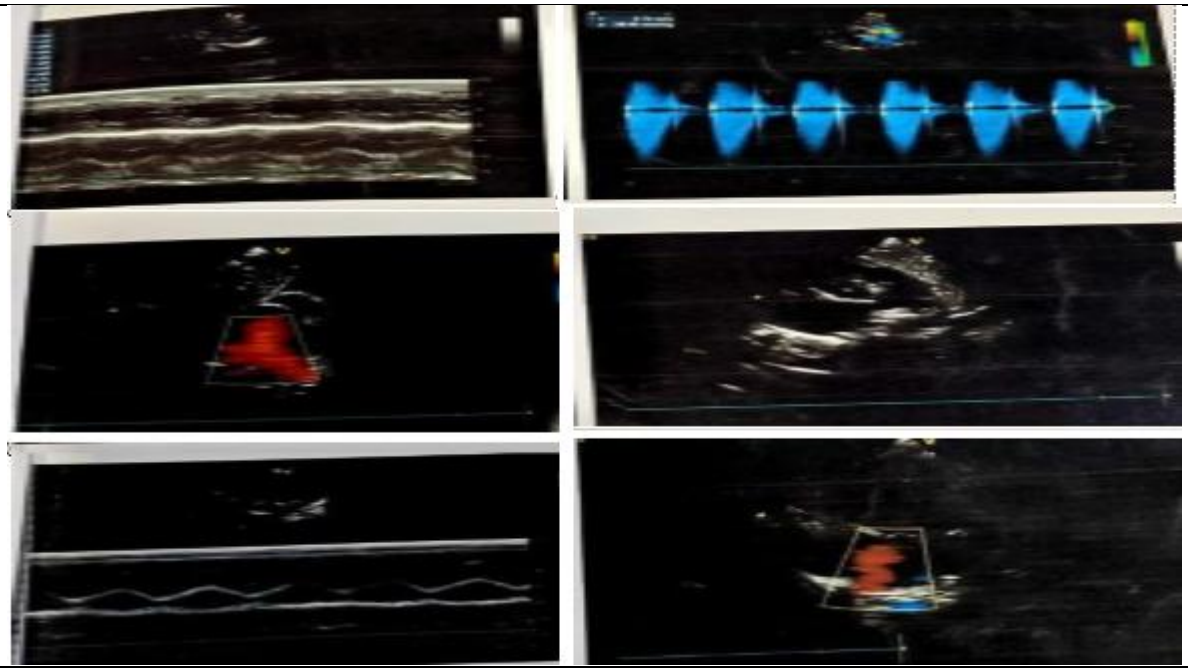

Figure 2: Doppler image across the pulmonary valve showing pulmonic valve PG 39.32 mmHg at 1 year of age

Challenges

1. Minor but painful surgery, vagal involvement.
2. Risk of bacterial endocarditis.
3. Sympathetic stimulation may increase pulmonary pressures, leading to hypoxia.
4. Opening up of small echo undemonstrable interchamber shunts leading to AV admixture.

Goals of Anaesthesia

1. No interference with cardiorespiratory pathophysiology.
2. Good pain relief.
3. High degree of suspicion and anticipation for shunt, hypoxia, hypercarbia, and acidosis.
4. Prophylaxis against endocarditis.
5. Pleasant anaesthesia experience in front of the mother, who was a healthcare provider.
6. Prevention of pressure and volume overload.

7. Optimal management of preload.

Conduct of Anesthesia Induction

The child was allowed his favourite civil dress and to be with his mother. He accepted a finger pulse oximeter. He was given a single intramuscular injection with ketamine 50mg, glycopyrrolate 0.1mg, and midazolam 1mg. The child's neck was supported by his mother, and friendly staff monitored his vital signs in the preoperative area.

After 5 minutes, fixed gaze developed, following which the child was wrapped in a towel and carried into a warm theatre and an operation table with a blow-over warmer. A 22 G intravenous cannula was inserted, under standard monitoring child turned left lateral while an assistant held an oxygen airway mask via JR circuit. Nitrous oxide was avoided. 100ml Ringer's lactate was started at 60ml/hr. A quick caudal block was administered with 0.25% Bupivacaine 10ml and 1% Lignocaine with Adrenaline 5ml. A Paracetamol 100mg suppository was placed rectally. Inj.ceftriaxone 500mg slow iv started for infective endocarditis prophylaxis. The child turned supine and was maintained on 02/Air and sevoflurane. The surgical site was checked for pain.

With absence of pain response on preputial pinch, surgery was proceeded with dorsal slit technique. The child was given an injection of morphine 1 mg intravenously for analgesia, Sedation, and pulmonary vasodilation.

With a stable Hemodynamics intra-operative period was uneventful, and the child had a stable and pleasant post-operative period.

DISCUSSION

Right-sided obstructive congenital heart disease is extremely variable in severity and presentation of signs and symptoms. Failure of tissue formation and coordinated growth leads to poor development of valvular structures and incomplete septation of the right ventricle. Impairment of normal hemodynamic blood pressure and flows from structural anomalies may further cause chamber dilation or hypertrophy, along with abnormal blood vessel growth and formation. The right-sided obstructive congenital heart diseases, in common occurrence after presenting with anaesthetic challenges, may be classified as: (a) Ebstein's anomaly, (b) Tetralogy of Fallot, (c) Pulmonary stenosis with intact ventricular septum (d) Pulmonary atresia with intact ventricular septum (e) Pulmonary atresia and VSD with major aortopulmonary collateral arteries.[1]

Pulmonary stenosis with an intact ventricular septum is a fairly common malformation, accounting for 10% of congenital heart defects. The lesions can be further divided into three groups: valvular, subvalvular, and supravalvular lesions. Valvular pulmonary stenosis is most common; about 80-90% of the reported cases are valvular, while subvalvular and supravalvular varieties are less common. Acute can range from asymptomatic mild valvular pulmonary stenosis to that of requiring urgent intervention, namely ductal-dependent or critical pulmonary stenosis. The stenotic pulmonary valve may be unicuspid, bicuspid, or tricuspid and is dome-shaped or dysplastic with a narrow central opening. Obstruction to RVOT may lead to hypertrophy of the infundibulum, eventually leading to diastolic dysfunction as ventricular compliance worsens. Valvular pulmonary stenosis is commonly associated with other defects such as Noonan syndrome, atrial septal defect, Ebstein's anomaly, double outlet Right ventricle, and transposition of great vessels.[2]

Subvalvular pulmonary stenosis can be divided into infundibular pulmonary stenosis and subinfundibular. Infundibular pulmonary stenosis is a part of tetralogy of Fallot, whereas sub infundibular pulmonary stenosis occurs within the context of a double-chambered right ventricle. Supravalvular pulmonary stenosis is seen in many disorders, such as congenital rubella, Williams, Alagille, DiGeorge, and Leopard syndrome.

Noonan syndrome is an uncommon autosomal dominant disorder with a prevalence of 1:1000 to 1:2000, characterised by dysmorphic facial features, hypertelorism, low-set ears, hypertrophic cardiomyopathy, pulmonic stenosis, mental retardation and bleeding disorders. Noonan syndrome, often called Male Turner Syndrome. The genes involved in this syndrome are components of the RAS/MAPK pathway, namely the mitogen- activated protein kinase Pathway.[3]

Embryology of Pulmonary Valves

The heart and vascular abnormalities are the largest category of human birth defects, accounting for 1% of malformations among live-born infants. The incidence among stillborn is 10 times as high. 8% of cardiac malformations are due to genetic factors, 2% are due to environmental agents, while others are multifactorial.

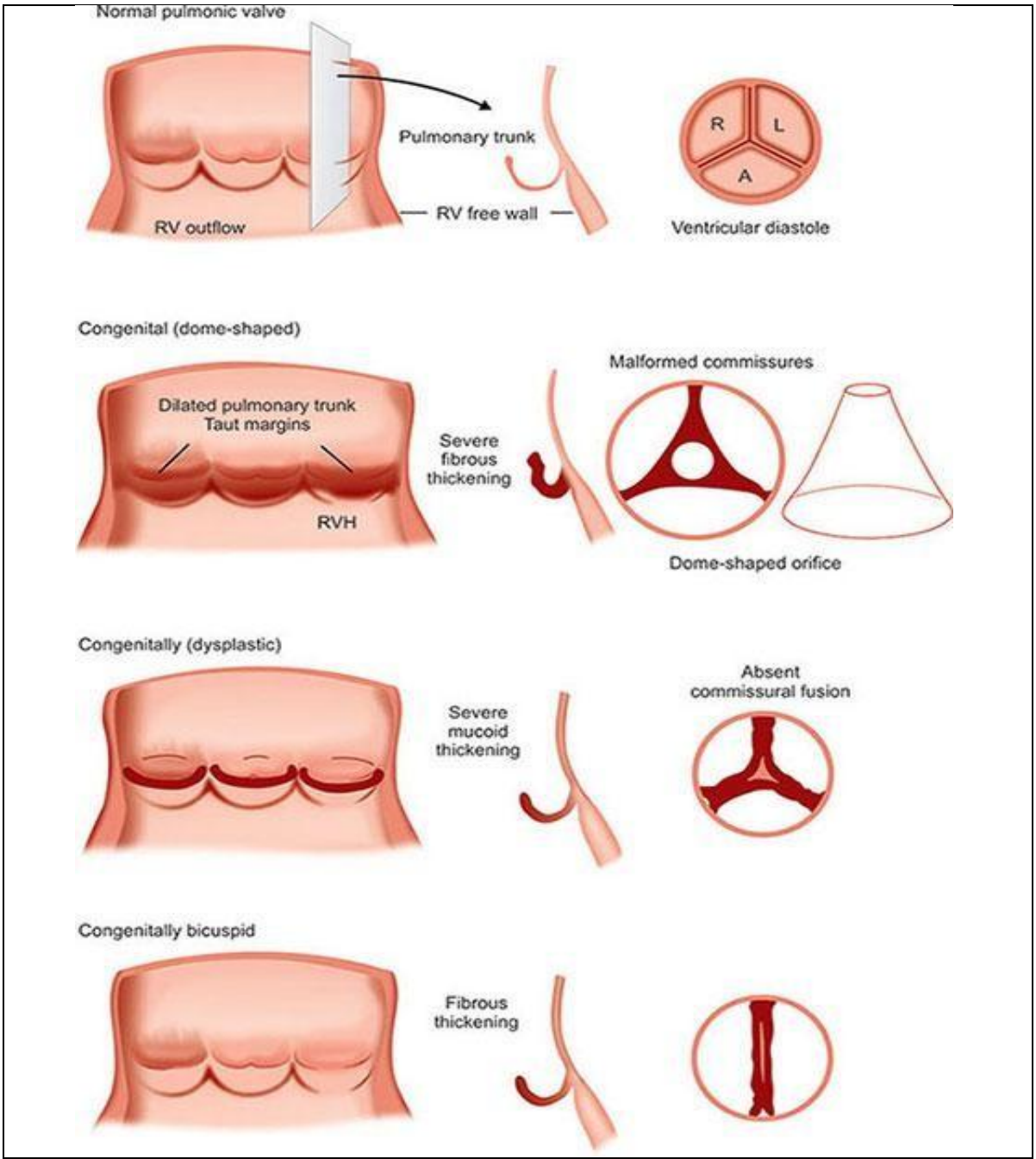

Figure 3: Anatomy of normal pulmonary valve and various congenital causes of pulmonary stenosis[4]

Septum Formation in the Truncus Arteriosus and the Conus Cordis

In the truncus, pairs of opposing ridges appear by the fifth week. These ridges, called truncus swellings or cushions, lie on the right superior wall, namely, the right superior truncus swelling, and on the left inferior wall. The right superior truncus swelling grows distally and to the right. Simultaneously, cushions develop along the right dorsal and left ventral walls of the conus cordis.

These swellings grow towards each other and distally to unite with the truncus septum. The fusion of the two conus swellings causes the septum to divide the conus into an anterolateral portion, which forms the right ventricular outflow tract, and a Posteromedial portion that forms the left ventricular outflow tract.[5]

The pulmonary valves develop from the Endocardial cushion in the outflow tract of the heart tube. These cushions undergo a process called Valvulogenesis, where they form the primordial heart valve. These cushions give rise to cusps of semilunar valves.

During late gestation and soon after birth, the valve cusps become stratified, developing into a fibrous tissue with collagen, proteoglycan and elastin. The cusps also develop their characteristic shape and structure, allowing them to open and close with each heartbeat.

The pulmonary valve leaflets have undergone endothelial-mesenchymal transition, become more organised and stratified, with the outer surface forming the ventricular aspect of the valve. The fibrous nodule and the pulmonary sinus also develop on each cusp.

Thus, the stages of formation of the pulmonary valve include endocardial cushion formation, valvulogenesis, pulmonary valve formation to separate the right ventricle and pulmonary trunk, delineation of the leaflet, maturation, and remodelling.

Histologically, the pulmonary valve consists of a stratified extracellular compartment of 4 layers, which are the arterialis, fibrosa, spongiosa and ventricularis. The arterialis faces the artery, and the ventricularis faces the ventricle.[6] Each of these layers has a different composition and function. The arterialis is the thinnest layer; the fibrosa is the backbone of the semilunar valves and consists of circumferentially arranged dense collagen networks and merges with the annulus and cardiac fibrous skeleton. The spongiosa layer is the layer that allows shear stress between layers during flexure and provides compressive strength because it consists of proteoglycans and glycosaminoglycans. The ventricularis layer contains most of the elastic fibres that assist with the elastic recoil of the cusps.[7]

The pulmonary valve opens at the systolic phase of the cardiac cycle, enabling the deoxygenated blood to be pumped from the right ventricle to the pulmonary circulation. It closes at the diastolic phase of the cardiac cycle, allowing sufficient filling of the right ventricle, the pulmonary valve at a diameter of about 20 mm.[8]

The pulmonary valve has no distinct blood supply or lymphatic drainage. The valve receives its innervation from branches of the cardiac ventricular plexus. The entire leaflet contains the nerve terminals except for the coaptating edge or the lower region compared to the aortic valve leaflet.

The pulmonary valve has more numerous nerve terminals. The activity of acetylcholine esterase or tyrosine hydroxylase, and neuropeptide Y is observable in these nerve terminals. With age, the density and distribution of innervation in the aortic valve leaflets decrease, but this does not occur in the pulmonary valve leaflets.[9]

Pathophysiology of Pulmonary Stenosis

Narrowing of the pulmonary valve leads to increased right ventricular workload, right ventricular hypertrophy, compromised cardiac output, eventually resulting in right ventricular failure, right atrial hypertrophy, arrhythmias, and pulmonary hypertension in some cases. Newborns with critical pulmonary stenosis may present with heart failure that results from suprasystemic right ventricular pressure, significant tricuspid regurgitation, and right-to-left shunt via atrial septal defect or patent foramen ovale. Individuals with mild to moderate pulmonary stenosis have been documented to survive into the 4th decade of life.

Patients with mild pulmonary stenosis have a normal electrocardiogram, whereas in severe stenosis, there may be right axis deviation and evidence of right ventricular hypertrophy. Echocardiography with Doppler evaluation can be used to grade mean and peak gradients, location of stenosis, presence of double-chamber right ventricle (DCRV), and estimate right ventricle volume and ejection.

Assessment of Pulmonary Stenosis

Echocardiography plays a major role in the assessment and management of pulmonary stenosis.[10] It is also essential in determining an appropriate management strategy.[11] Ancillary findings with pulmonary stenosis, such as right ventricular hypertrophy, may also be detected and assessed. Although the majority of pulmonary stenosis is valvular, narrowing of the RVOT below the valve from concurrent right ventricular hypertrophy may occur, as there may be narrowing of the pulmonary artery sinotubular junction above the valve.

Grading of Pulmonary Stenosis

Quantitative assessment of pulmonary stenosis severity is based mainly on transpulmonary pressure gradient (TPPG). Calculation of pulmonic valve area by planimetry is not possible since the required image plane is, in general, not available. The continuity equation, are proximal isovelocity surface area method, has not been validated in pulmonary stenosis. The estimation of systolic pressure gradient is derived from the transpulmonary velocity flow curve using the simplified Bernoulli equation ($\Delta P = 4V^2$).

In most instances of valvular pulmonary stenosis, the modified Bernoulli equation works well, and there is no need to account for the proximal velocity as this is <1m/s.

Pulsed wave Doppler may be useful to detect the sites of varying levels of obstruction in the outflow tract, and in less degrees of obstruction, may allow a full evaluation. Muscular infundibular obstruction is characterised by a late peaking systolic jet that appears dagger-shaped. This pattern is useful in separating dynamic muscular obstruction from fixed valvular obstruction, where peak velocity is generated early in systole. In pulmonary valve stenosis, the pressure gradient across the valve is used to ascertain the severity of the lesion more so than in left-sided valve conditions, due to difficulty in obtaining an accurate assessment of the pulmonary valve area. The following definition of severity has been defined in the 2006 American College of Cardiology / AHA guidelines on the management of valvular heart disease.[12]

Nerve Supply of Penis

The penis is innervated primarily through pudendal and cavernous nerves from the sacral spinal cord segments S2, S3, and S4. Somatic sensory and motor/innervation provided by the pudendal nerve, while the cavernous nerves provide parasympathetic and visceral afferent fibres involved in erection. {Figure 4}

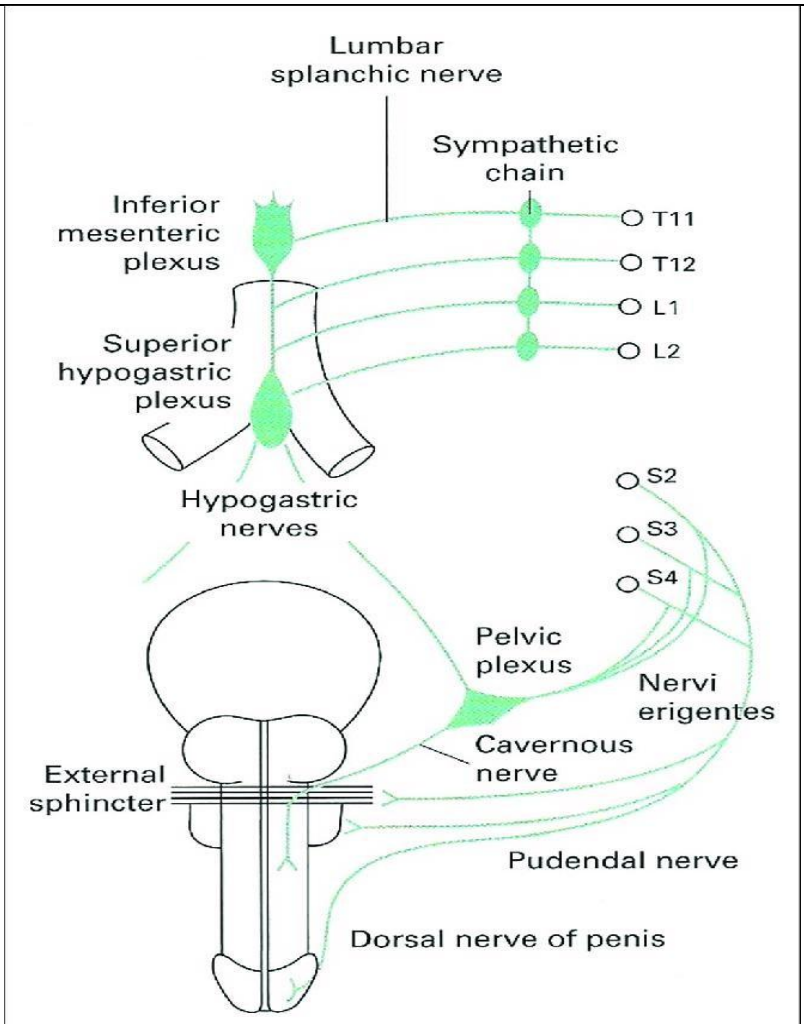

Figure 4: The nerve supply of the penis. From Eardley and Sethia[13]

Pudendal Nerve
Root value: S2 S3 S4
Branches: Dorsal Penile Nerve carries Sensory information from glans and shaft of penis including frenulum. It also provides somatic motor innervation to the bulbocavernosus muscle.

Cavernous Nerves: These are derived from pelvic splanchnic nerves that supply erectile tissue with parasympathetic fibres; these are crucial for erection.

Ilioinguinal Nerve: Innervation of the root of Penis.

Sensory Nerve Endings: Contribute to sensitivity of the glans penis.

Autonomic Supply: Sympathetic and parasympathetic supply regulate the blood flow and muscle tone of the penis. Helicine arteries, branches of deep penile artery and venous drainage by sub truncal venular plexus form the blood supply to penis.

Anaesthesia for Circumcision
Though a simple appearing surgical procedure, a comprehensive analgesia and anaesthesia is needed due to multiple innervations of penis. Nerve blocks, including the dorsal penile nerve block and a ring block at the root of the penis themselves need sedation and analgesia in a child for successful performance. The dorsal penile nerve is the major somato sensory nerve supply to the penis as it runs ventrolaterally from glans to frenulum. The perineal branch of pudental nerve also supplies

frenulum which is the most painful and vascular part of the penis. This is missed in the dorsal penile nerve block. Hence, a caudal epidural block is the most comprehensive definitive mode of anaesthesia with effective postoperative analgesia.[14]
Which, with 0.25% bupivacaine as 1ml/kg in less than 15kg children, allows a pain-free passage of the first urine by the child after surgery. Motor block to the lower limbs is minimal, and the child usually can kick their feet on the OT table itself.

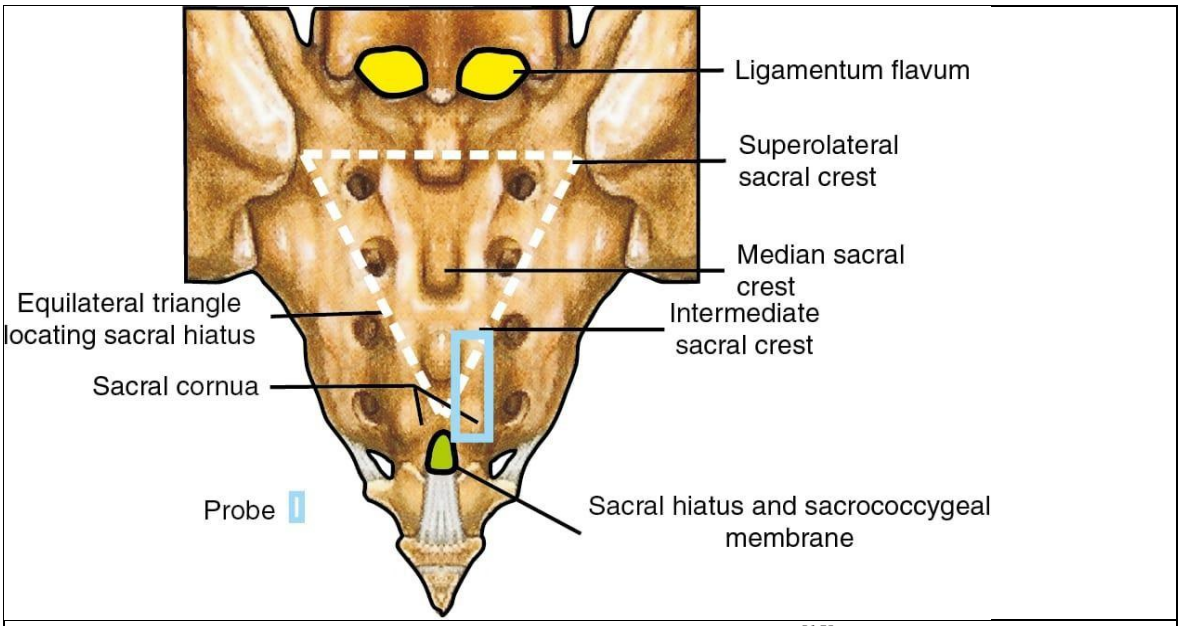

Figure 5: Anatomy of sacral hiatus[15]

CONCLUSION

This case report of anaesthesia for circumcision in a child with acyanotic congenital heart disease, with a high degree of suspicion of valvular pulmonary stenosis under periodic surveillance, was safely done under caudal analgesia without interfering with the cardiovascular physiology of the child. The approach to sedate and perform the block offers a major contribution to allay sympathetic and parasympathetic stimulation that would precipitate laryngospasm or increased pulmonary pressures in children with cardiovascular compromise.

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