CASE REPORT

Cardiac Failure and PPHN due to extra cardiac cause in newborn (Vein of Galen malformation)

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A 6 hours old male child born to primi mother out of non-consanguinous marriage. Baby cried immediately after birth and APGAR score was good. Birth weight 3kgs. Noted mild respiratory distress at birth, admitted in the NICU and given supplemental Oxygen and IVFs. Gradually respiratory distress increased and noted grunting, increasing precordial pulsations, SPO\textsubscript{2} fluctuation and a cardiac murmur. Chest X-ray was taken and it revealed Cardiomegaly. Referred to us for further management.

On admission noted grunting, tachypnoea, sternal, subcostal and intercostal retractions, mild precordial and epigastric pulsations, HR:188/min., RR 110/min., SPO\textsubscript{2} 80-85% fluctuating, BP:40/30/38 mmHg, CFT:4 secs., all peripheral pulses felt, but feeble, colour appears to be pale. With nasal prong Oxygen, SPO\textsubscript{2} improved to 92%. On systemic examination: CVS: S\textsubscript{1} S\textsubscript{2} heard, loud P\textsubscript{2} present and systolic murmur of grade 4/6 was present. Respiratory system: respiratory distress present and bilateral air entry was equal. Abdominal examination: Right hypochondriac fullness present, liver enlarged 4 cm below the RCM. CNS examination; normal AF and irritability present, tone and reflexes were normal.

Taken chest X-ray and it revealed cardiomegaly. Echocardiography revealed severe pulmonary artery hypertension, tricuspid regurgitation, enlargement of right atrium and right ventricle, dilatation of great vessels and no structural cardiac abnormalities. Taken blood for all septic work-up. At this present clinical scenario with normal hematological findings (Hb:17.9g%, TC:17,000/mm\textsuperscript{3}, DC: P-78%, L-19%, E-3%, platlets:260000/mm\textsuperscript{3}), we started nasal prong Oxygen, Digoxin, furosemide, fluid restriction and inotrope (Dopamine). Gradually over 6 hours baby started passing urine, SPO\textsubscript{2} increased to 96% and heart rate came down to 170/min. On 2\textsuperscript{nd} day, the calculated urine output was <1ml/kg/hr and also noted mild haematuria, jaundice (serum bilirubin Total:10mg/dl and direct: 1mg/dl), liver 5 cm below the RCM, mild abdominal distension, HR:160-170/min, RR:80-90/min with increase indrawing, CFT:>4 secs, BP:45/35/42 mmHg, SPO\textsubscript{2} 89-90% fluctuating, increase in precordial pulsations and colour appears to be pale. Taken second opinion for the cardiac status and the reports were same (ECHO). Placed the baby on CPAP 5cm with 80% Oxygenation and SPO\textsubscript{2} stabilized to >95%. Continued the same treatment along with phototherapy for 2 days. On day 4, the urine output was 0.9ml/kg/hr, urea:68mg/dl, creatinine:1.8mg/dl. Serum bilirubin:14mg/dl, baby’s irritability increased, perspiration noted, indrawing increased, murmur was of same intensity, liver:5.5cm below the RCM, all peripheral pulses were bounding, neck pulsations were prominent and mild bleeding from the nasogastric tube noted. Repeat chest X-ray was taken and it revealed cardiomegaly with mild increase in pulmonary markings. With all these features
we suspected extra cardiac problem for the cardiac failure. with high index of suspicion of. A-V malformation/ aneurysm the cranium examintion revealed Bruit. Done the bedside cranial ultrasound which showed huge vein of Galen aneurysm with shifting of the right lateral ventricles without hydrocephalus. At the same time, ultrasound abdomen revealed hepatospleenomegaly, ascites and gall bladder congestion which were suggestive of congestive heart failure. For more confirmation requested MRI brain scan with Venogram which revealed huge vein of Galen aneurysm / malformation with multiple feeders (Choroidal in type). Later taken Neurosurgeon’s opinion. After collective decision about baby’s poor prognosis and financial burden for parents so decided to withdraw the treatment.

**Discussion:** Vein of Galen malformation is not a true aneurysm. It is only a artiio-venous fistula (AVF) between deep choroidal arteries and the median prosencephalic vein (MPV) of Markowski. Because of this fistula, vein of Galen is not able to form an aneurysm.It is only a pooling site(Pouch).

Abnormal connection of choroidal arteries to median prosencephalic vein (MPV) occurs at 6-11 weeks of gestation. Flow through this fistula prevents normal regression of MPV, so multiple abnormal connections will develop. It is sporadic in occurance and there is no increased risk to siblings. Male: female ratio is 2:1.It is most commonly diagnosed in the newborn period and rarely after 3 years. If diagnosed in older children or adults, it is usually compensated. It comprises less than 1% of cerebral vascular malformations at any age, but in the paediatric vascular malformation group it might account upto 30%. It is the most common extra cardiac cause of high output cardiac failure in newborns. Sometimes it may be associated with ASD(may exacerbate CHF) and aortic coarctation. Probable causes includes (assumption): a) Venous occlusion/ stenosis leading to increased flow and pressure. b) Cerebral ischemia/atrophy leading to arterial steal and /or chronic venous hypertension. c) Hydrocephalus due to decreased CSF resorption or cerebral aqueductal obstruction.

Sometimes it might cause malformation of adjacent brain structures like Penial gland and 3rd ventricle. Microscopically wall of the venous pouch is thickened. The classification (based on angio-architecture): 1.Choroidal type has multiple feeders from pericallosal, choroidal and thalamo perforating arteries. 2.Mural type: has single or few feeders from colllycular or posterior choroidal arteries.

Clinically most common signs/symptoms: a) in Neonate: high-output cardiac failure and cranial bruit.. b)In infants and older children: Macrocrania, prominent superficial cranial veins, developmental delay, failure to thrive, hydrocephalus, seizures, headache and hepatic failure.

**Imaging findings: Ultrasound:** a)Prenatal studies can identify malformation in 2nd and 3rd trimester.Cardiac dilatation and hydrops foetalis carries poor prognosis. b)Postnataally it is useful for early bedside diagnosis ,provided there is large midline varix located in quadrigeriminal plate cisterna. Size can vary upto several centimeters. Morphologically it might look spherical / tubular varix.

**Echocardiography:** Shows dilatation of right heart, SVC and ascending aorta, severe pulmonary artery hypertension and PDA with significant R-L shunt (poor prognosis).
**MRI findings:** Fetal MR can identify malformation along with absence of brain or other end organ injury (significant antenatal end organ injury is a contraindication to aggressive treatment). MRA (magnetic resonance angiography) study is the key for pre treatment assessment of feeders to malformation (MRA is unaffected by coils or acrylic embolic material). MRV (magnetic resonance venogram) is essential in initial and follow-up evaluation (presents and degree of venous stenosis can have major influence on prognosis). Varix contents are hypointense and homogenous (T1 signal), sharp delineation of malformation and prominent flow voids from feeding arteries around varix (T2 signal).

Prognosis is related to volume of shunt and timing/success of treatment. High volume shunts requiring treatment in newborn period have worst prognosis. Delay in treatment until 4-6 months is associated with better outcome. Without treatment, progression of CHF or brain damage results in death. Up to 60% will be neurologically normal after treatment.

**Treatment:** Intractable CHF, multi organ failure and brain damage at presentation are contraindication to treatment.

Medical therapy for CHF until 4-6 months (failure of therapy warrants earlier neuro-intervention.).

Trans catheter embolisation (TCE) at 4-6 months is the best available treatment at present. Permenent occlusion of fistula point from arterial side is done. May require several stages of embolisation (upto 5-7 stages). Filling of venous pouch is less effective. Frequent neurological and MRI assessment after TCE is needed. If deterioration is noted, further embolisations are needed. Treatment for hydrocephalus is controversial and is reserved for refractory hydrocephacus after all TCEs performed. Because shunt placement is associated with exacerbation of venous ischaemia and there is risk of haemorrhage from enogorged subependymal veins. Over all these procedures are cost effective (3-5 lakhs).

**Conclusion:** In our case, baby had high volume shunt and progressive multi organ failure, poor prognostic indicators were present, and also in view of financial aspects parents were not willing to continue the treatment and baby was discharged at request.

**References:**
1. Rennie JM, Roberton NRC. CNS malformations, Textbook of Neonatology 3rd edn ; 1309.
Precordial bulge & Cardiomegaly
Right hypochondriac fullness
Tall & wide P waves

Tricuspid Regurgitation
D shaped Left ventricle Edematous gall bladder & ascites

Doppler study of vein of Galen & markings in u/s brain
M R I picture of the same

M R I venogram showing malformation and feeding arteries