Objective:

To describe the clinical and echocardiographic features and management of thiamine-responsive acute pulmonary hypertension (TRAPH). Design: Prospective observational study: January 2013 to November 2019.

Patients:

All exclusively breastfeeding infants with severe Pulmonary hypertension (PH) diagnosed by ECHO were recruited. Interventions: Intra- venous thiamine 100 mg diluted in 10 mL of normal saline was given as an infusion over 1hour, once a day for 3 days for all babies.

Main outcome measures:

Clinical resolution of heart failure and normalisation of PH by echocardiogram.



ACADEMIC P.E.A.R.L.S

Pediatric Evidence And Research Learning Snippet



Thiamine Responsive Acute Pulmonary Hypertension

Thiamine-responsive acute severe pulmonary hypertension in exclusively breastfeeding infants: a prospective observational study. Arch Dis Child 2020;0:1-6.

Results:

- A total of 250 infants had severe PH and 231 infants responded to thiamine.
- Clinical improvement with complete resolution of PH was noticed within 24–48 hours. PH completely resolved in 92% (231/250) after thiamine administration
- Babies were followed up to a maximum of 60 months with no recurrence of PH.
- Echocardiogram showed reduction in chamber dimensions ,resolution of TR and left-to-right shunting across PFO within 24 to 48 hours.

Conclusion:

Thiamine deficiency is still prevalent in selected parts of India. It can cause lifethreatening PH in exclusively breastfeeding infants. Thiamine administration based on clinical suspicion leads to remarkable recovery.

Key Message:

- Thiamine deficiency can present as acute pulmonary hypertension in exclusive breast feeding infants.
- The mean reduction in PASP from admission to discharge was 52.1mmHg (95%Cl 50.2 to 53.9) (reduction of 71%) with p value of <0.01 following administration of Thiamine

EXPERT COMMENT



"Thiamine Responsive Acute Pulmonary Hypertension (TRAPH) can be considered as differential in exclusively breast fed infants presenting as acute right heart failure"

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Hon. Secretary Gen. 2020-21

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<u>Reference</u>

Sastry UMK, M J, Kumar RK, et al.Thiamine-responsive acute severe pulmonary hypertension in exclusively breastfeeding infants: a prospective observational study. Archives of Disease in Childhood Published Online First: 03 September 2020. doi: 10.1136/archdischild-2019-210777