

Recurrent non-immune hydrops improves with incidental maternal beta-blocker therapy

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INTRODUCTION

We present a case of recurrent non-immune hydrops (NIH) associated with a novel gene variant. Hydrops was treated with shunting and ultimately responded to propranolol initiated for maternal SVT.

CASE DESCRIPTION

- 35yo G6P3113
 - G1-3 Term SVD, no complications
 - G4 Neonatal demise at 27wk after pleural effusions and hydrops, failed shunting and Preterm labor
- 14wk with Cystic Hygroma (**A**)
- 17+5 Normal survey, Low risk NIPT
- 25+2 Bilateral effusions (**B**)
- 26+1 Thoracentesis of chylothorax. Maternal sirolimus therapy initiated.
- 27+3 Bilateral shunts placed (**C**)
- Sirolimus stopped due to maternal hepatotoxicity (resolved after 6d)
- 29+5 Mother presented in SVT at rate 170bpm. Cardioversion then on propranolol 40mg PO BID
- 30+5 Pelvic pain, noted large theca lutein cysts (**D**). Managed pain, resolved with hydrops
- Repeat CS at 36+1 w/o complication
- Neonate treated with propranolol and diuretics (weaned off). MRI: prominent lymphatic ducts. Managed conservatively, discharged at 12wk of life, and healthy without further treatment at 2 ys of age

TEACHING POINTS

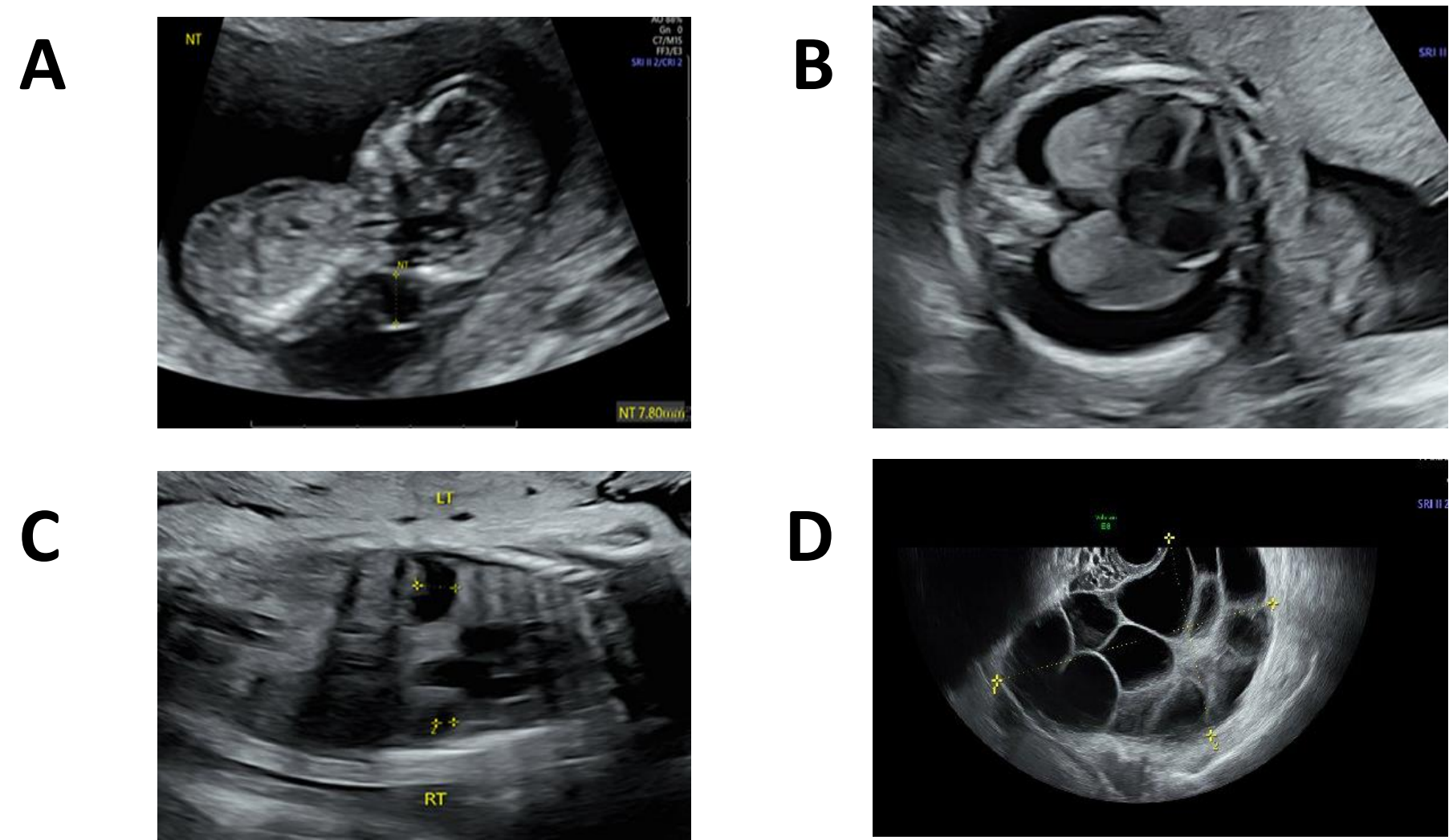
1. **Chylothorax (CT) accounts for 5-6% NIH**
2. **Theca Lutein Cysts associated with NIH and excess HCG production**
3. **Identified Novel EPHB4 variant**
 - A. Autosomal Dominant, in mother and 2 affected offspring, previously “VUS”
 - B. Variable expressivity and reduced penetrance
 - C. Leads to loss of phosphorylation ERK and impacts lymphatic specification
 - D. Other variants associated with lymphatic malformations
4. **Treatment of Chylothorax (Neonatal)**
 - A. Conservative – Nutritional management (MCT formula or TPN)
 - B. Surgical
 - ☐ Drainage
 - ☐ Thoracic Duct Ligation/Embolization
 - ☐ Pleurodesis
 - C. Medical –*variable results and side effects*
 - ☐ Octerotide (Acromegaly, Tumors)
 - ☐ Sildenafil (Pulmonary HTN)
 - ☐ Sirolimus (Rhabdomyoma)
 - ☐ **Propranolol**
5. **Propranolol use prenatally**
 - A. Based on use for Neonatal CT ⁴
 - ☐ Post-op CT after congenital cardiac surgery ²
 - Reduced chest tube days, infection and LOS
 - B. **Prenatal use** ¹
 - ☐ **40mg PO QID → resolution avg 35d**
 - ☐ **Delivered 37 and 39wk vs. 34wk**
 - ☐ **Shorter LOS, fewer readmission and interventions**
 - ☐ **No maternal or neonatal complications**

CONCLUSION



- Inadvertent use of propranolol for maternal SVT led to resolution of hydrops, and prolonged pregnancy with excellent neonatal outcome
- More studies on prenatal use of propranolol are warranted
- Identified novel, disease- causing EPHB4 variant

IMAGES



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