

## TEACHING FILE

## GRAND ROUNDS

Ira Shah

**JAUNDICE AND PULMONARY STENOSIS**

**Clinical Problem:-** A 2½ years old child born of third degree consanguineous marriage presented with gradual abdominal distension and jaundice since 1½ months. She was seen by a physician 1½ months back and treated with steroids and also received a blood transfusion. There is high coloured urine. Milestones are normal and birth was uneventful. She is immunized till date but has not received Hepatitis B vaccine. On examination, she has steroid facies, is normal nourished, has jaundice, hepatosplenomegaly and other systems are normal. Investigations showed:

- Bilirubin = 11.5 mg/dl (Direct bilirubin = 5.8 mg/dl)
- SGOT = 118 IU/L, SGPT = 120 IU/L
- Total proteins = 5.9 gm/dl, Albumin = 3.4 gm/dl
- Serum alkaline phosphatase = 553 IU/L (Normal)
- Prothrombin Time and partial thromboplastin time = Normal
- USG Abdomen = Hepatosplenomegaly. No varices
- Liver biopsy = Bile duct paucity
- Echocardiography = Left ventricular hypertrophy with mild pulmonary stenosis
- CBC = Normal
- HIV/ HBsAg = Negative

**What is the diagnosis ?**

**Expert's opinion:-** This child has arteriohepatic dysplasia (hepatic – bile duct paucity & arterial – pulmonary stenosis) which is also known as syndromic bile duct paucity or Alagille syndrome. It consists of a paucity of interlobular bile ducts with chronic cholestasis. Patients with this syndrome typically have the following major features: peculiar facies, chronic cholestasis, butterfly-like vertebral arch defects, and peripheral pulmonary artery hypoplasia or stenosis; either isolated or associated with complex cardiovascular abnormalities.

**DOI:** 10.7199/ped.oncall.2012.66

**CYTOMEGALOVIRUS INFECTION AND HEPATITIS**

**Clinical Problem:** - A 4 month old boy, 2nd born of third degree consanguineous marriage presented with not gaining weight since birth. He was a full term normal delivery with birth weight of 3 kg. Mother had fever at 7-8 months of gestation and had received antibiotics orally for same. The child on Day 15 of life developed rash over body with fever which cleared off in 4 days. He was given IV antibiotics for 4 days. He had recurrent cough since 1½ months of age with regurgitation of food since 15 days for which he was advised propped up position. His milestones and immunization were normal. He was on breast

feeds and started on formula feeds, a month back in view of poor weight gain. At 2 months of age, had hematemesis with malena and was given whole blood transfusion and fresh frozen plasma. An older male child had died at 1½ months of age due to failure to thrive. On examination, he was malnourished (weight = 2.75 kg, length = 56 cm), had pallor, large ears and firm hepatomegaly. Other systems were normal. Investigations showed:

- Hemoglobin = 10.8 gm/dl
- WBC count = 27,300/cumm (78 percent polymorphs, 20 percent lymphocytes)
- Platelet count = 8,07,000/cumm
- Venous blood gas = pH = 7.362, bicarbonate = 21 mmol/L
- Urine = Normal
- CRP = Negative
- Bilirubin = 2.5 mg percent (direct = 1.3 mg percent)
- SGOT = 123 IU/L, SGPT = 66 IU/L (elevated)
- Total proteins = 6.7 gm percent, albumin = 2.3 gm percent
- S. creatinine, electrolytes, calcium, alkaline phosphatase = Normal
- VDRL, HIV ELISA = Negative
- Ultrasound abdomen = Normal
- Urine for reducing substance = Negative
- TORCH = Cytomegalovirus (CMV) IgG positive, Rest all IgM and IgG negative
- Blood culture = No growth
- CMV viral load = 3000 copies/ml
- Hearing and eye examination = Normal
- GER = Normal study

After 10 days, liver enzymes and bilirubin were normal and CBC was also normal and child's regurgitation episodes also decreased.

**Does the CMV infection require treatment?**

**Expert's opinion:-** This child has a cytomegalovirus (CMV) disease whether congenital or acquired (due to blood product transfusion) is not known. However, he had liver dysfunction which has recovered on its own. His hearing and eye evaluation are also normal. Also there is no pneumonia. Thus active CMV organ dysfunction is not found in the child. Though CMV viral load is 3000 copies/ml it is not that high to warrant treatment especially since child does not have symptomatic CMV disease. CMV is going to remain in the body for lifetime and will cause problems only when the immunity goes down and CMV flares up. The drugs available to treat CMV are all which will arrest the viral replication but will not cure the disease. Also the drugs for treating CMV such as ganciclovir, valganciclovir foscarnet are all toxic and can lead to severe adverse effects. Hence treatment is recommended only when there is symptomatic CMV disease and viral load is a

method by which one can tailor the treatment. (Therapy to be continued till viral load becomes undetectable).

This child needs annual hearing and fundus evaluation for early screening of CMV induced deafness and chorioretinitis respectively.

**DOI:** 10.7199/ped.oncall.2012.69

**EXPOSURE TO XDR-TB**

**Clinical Problem:-** A 3½ years old girl is in contact with an adult suffering from XDR-TB. Her Mantoux test is also positive. She is otherwise asymptomatic. Her Chest X-Ray is normal.

**How should this child be treated?**

**Expert's opinion:-** This child has latent TB, is less than 5 years of age and is in contact with an adult having TB. Thus this child should receive prophylaxis for TB. As per WHO, prophylaxis for latent TB is 6 months of INH. However, this child is in contact with an adult having XDR-TB. Thus she may have been exposed to resistant bug. Thus INH may not be protective. There is not enough literature to determine which drug



should be given as prophylaxis in such a scenario. WHO recommends a wait and watch policy in such a situation and to treat the child only when the child becomes symptomatic. There are reports of use of pyrazinamide and ofloxacin prophylaxis in such situations but it is not recommended as you may actually lead to more drug resistance in the child.

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**From:** Medical Sciences Department, Pediatric Oncall, Mumbai.

**Address for Correspondence** Dr Ira Shah, 1/B Saguna, 271, B St Francis Road, Vile Parle (W), Mumbai 400056.

**E-published:** December 2012 . **Art#**75

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**DOI:** 10.7199/ped.oncall.2012.75

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