Management of intractable pruritus in a child with cholestatic liver disorder
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BACKGROUND
Childhood cholestatic hepatobiliary disorders present with pruritus which is bothersome and difficult to manage. The pathogenesis of pruritus in cholestasis is unknown but several hypotheses have been proposed, including bile acid accumulation, increased opioidergic tone and elevations in lysophosphatidic acid levels. Pruritus can have a profound effect on the patient’s quality of life resulting in sleep deprivation and emotional disturbances. Treatment of the primary cause of cholestasis is important to address the pruritus. However, symptomatic treatment is often necessary especially in cases where the underlying hepatobiliary disease cannot be corrected.

CASE DESCRIPTION
Currently, 19-month-old male was first seen at the gastroenterology clinic at age of 5 months. He was referred as a case of prolonged conjugated hyperbilirubinaemia. He was born at term with birth-weight (BW) of 2kg. He had a history of jaundice for 2 weeks. There was no pale stools or dark urine and has doubled his BW.

On examination, he was jaundiced, had no dysmorphic features, but developed the typical facies of Alagille syndrome over time. Investigations showed conjugated hyperbilirubinemia with raised ALP, GGT and transaminases. A liver biopsy showed severe paucity of bile ducts in 12 portal areas.

2 weeks after the initial visit he presented with severe generalized itching associated with bleeding upon scratching, irritability and poor sleep. A presumptive diagnosis of cholestasis-associated pruritus was made after any possible dermatologic disorder associated with pruritus were ruled out.

Ursodeoxycholic acid was started with no improvement even at a maximum dose of the medication. Phenobarbitone and rifampicin were added in a stepwise manner which improved pruritus, but there were challenges with obtaining rifampicin after 9 months of treatment hence cholestyramine was added to the ursodeoxycholic acid and phenobarbital, but the child’s pruritus worsened. Though clear instructions were given to the mother regarding administration of cholestyramine we thought the worsening of the child’s pruritus was still possibly due to chelation of the other medications by cholestyramine, hence it was suspended.

An antihistamine, cetirizine was added to child’s treatment and his mother reported some improvement in pruritus but this was short-lived. Finally parenteral infusion of naloxone, an opioid antagonist, was administered over 24 hours with significant improvement in child’s symptoms like scratching, irritability and sleeplessness.

Child is currently on oral ursodeoxycholic acid, naltrexone, phenobarbitone and cetirizine with continued remarkable response.

CONCLUSION
Pharmacologic treatment with recommended medications in a stepwise approach is essential for symptomatic relief of the pruritus.