stored for future analysis in definitive diagnosis. Crigler-Najjar syndrome type I was considered in our case by its classical clinical presentation, non-response to two trials of phenobarbitone therapy and by a process of exclusion of other conditions causing persistent unconjugated hyperbilirubinermias in infancy. Currently the treatment of CNS-I consists of phototherapy followed by liver transplantation(2), Tin-mesoporphyrin, hepatocyte transplantations(5), ursodeoxy-cholic acid, bilirubin oxidase, antioxidants, calcium supplements, clofibrate, flumecinol, chlorpromazine and urine alkalinization have been tried in CNS-I. The most exiting new therapy discussed is the gene therapy(5).

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Improving Oxygenation in Preterm Neonates with Respiratory Distress

We read with interest the communication by Patole, *et al.*(1). Definitely the adminis-tration of antenatal glucocorticoids and availability of Hudson-Prongs (HP) conti-nuous positive airway pressure(CPAP) are remarkable breakthroughs in management of preterm babies and have played a role in reducing the need for ventilation. Our neonatal set up is very similar to the authors and we would like to share our experience of a similar case. This baby was the second of twins delivered by cesarean section in view of fetal decelerations at 30 week gestation and weighed 1245 grams. He was transferred from operation theater to newborn care with CPAP support by Neopuff resuscitaire and was put on 5 cm CPAP in 28% FiO_2 (fraction of oxygen in inspired air). Chest radiograph at six hours of life was typical of hyaline membrane disease. At about 24 hours of birth, he deteriorated and oxygen requirement rose to CPAP of 6 cm with 40% FiO_2 and upto 55% in next four hours. We were in a dilemma at

this stage-whether to initiate ventilation and give surfactant or give a watchful trial of 7 cm CPAP. After transillumination for possible pneumothorax was negative, we chose the latter option. The other measures adopted included using bigger prongs (so as to have a more snug fit and avoid leak), keeping in prone position and putting a rolled soft towel under neck. The former helps by better aeration of dependent regions of the lungs thus improving ventilation/perfusion ratios(2) and the latter by splinting the upper airway. Within three hours FiO_2 could be weaned to 35% and CPAP to 5 cm, thus obviating the need for ventilation. This improvement was sustained and the baby could be rapidly weaned off oxygen support. CPAP is a critical component of therapy in RDS where loss of surface area is the main denominator and it helps by improving alveolar recruitment. Currently, a multicentric trial (coordinated from Melbourne) is underway in Australia, randomizing infants to CPAP or intubation, the primary outcome being the incidence of Chronic Lung Disease. What would be of interest is to identify variables, which will help recognize babies likely to fail trial of CPAP, so that ventilation could be initiated in them straight away.

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Distal Asymmetric Spinal Muscular Atrophy Involving Upper Limbs

I read with interest the recent article by Mishra, *et al.*(1). They describe an interesting case of distal asymmetric spinal muscular atrophy involving upper limbs associated with high serum lead levels. However, I would like to make certain comments.

Firstly, the case presented here has several features against a diagnosis of Hirayama disease:

1. *One year duration of symptoms:* A diagnosis of Hirayama disease is typically made after at least four to five years of symptoms(2) (after demonstrating a stationary clinical course following a progressive disease during initial four to five years of illness),
2. *Symmetrical onset of symptoms:* Hirayama disease is characterized by a unilateral-predominant involvement of upper limbs. Though a bilateral upper limb involvement is seen in about 20% of cases, it is highly asymmetric, showing a peculiar "oblique atrophy"(3),
3. *Female sex:* Hirayama disease is ten times more common in males(2),
4. *Age of six and a half years:* The commonest age of onset is between 15-25 years and onset below 10 years of age has