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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REFERENCES

1. Patole SK, Mcglone L, Hares D. Improving oxygenation in preterm neonates with respiratory distress. Indian Pediatr 2002; 40: 376.
2. Martin RJ, Herrel N, Rubin D, Fanaroff A. Effect of supine and prone positions on arterial oxygen tension in the preterm infant. Pediatrics 1979; 63: 528-531.

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

not been reported before.

5. *Normal MRI of the spine:* MRI abnormalities are almost universal. In an Indian study, focal cord atrophy was seen in 100% of cases at the level of cervical 4 to 7 vertebral bodies in MRI done in neutral neck position(4).

Secondly, I disagree with the inclusion of Madras motor neuron disease (MMND) as a differential diagnosis in this case. MMND typically presents with the involvement of lower cranial nerves (seventh and ninth to twelfth) and sensorineural deafness along with features of chronic anterior horn cell disease(5,6).

The diagnosis in this child could still be a genetically determined distal spinal muscular atrophy. Genetic and molecular studies to identify survival motor neuron (SMN) 1 and 2 gene mutations would have been useful towards making a definite diagnosis(7).

Finally, the association of elevated serum lead level in this child appears to be coincidental. However, chelating therapy is required for the same.

In conclusion, the diagnosis of Hirayama disease cannot be made confidently in this case. Genetic studies would have been useful in making a definite diagnosis of spinal muscular atrophy.

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REFERENCES

1. Mishra D, Agrawal A and Gupta VK. Distal spinal muscular atrophy of upper limb (Hirayama disease) associatemd with high serum lead levels. Indian Pediatr 2003; 40: 780-783
2. Gouri-Devi M, Nalini A. Long-term follow-up of 44 patients with brachial monomelic amyotrophy. Acta Neurol Scand 2003: 107: 215-220.
3. Hirayama K. Juvenile muscular atrophy of the distal upper limb–three decades of description and it's treatment. Rinsho Shinkeigaku 1993; 33: 1235-1243.
4. Pradhan S, Gupta RK. Magnetic resonance imaging in juvenile asymmetric segmental spinal muscular atrophy. J Neurol Sci 1997; 146: 133-138.
5. Meenakshisundaram E, Jagannathan K, Ramamurthi B. Clinical pattern of motor neuron disease seen in younger age groups in Madras. Neurol India 1970; 18: Suppl 1: 109-112.
6. Gourie-Devi M, Nalini A. Madras motor neuron disease variant, clinical features of seven patients. J Neurol Sci 2003; 209: 13-17.
7. Panigrahi I, Kesari A, Phadke SR, Mittal B. Clinical and molecular diagnosis of spinal muscular atrophy. Neurol lndia 2002; 50: 117-122.

Reply

Our comments are as under:

1. None of the previous authors have used the criteria of 'four to five years of symptoms' before making a diagnosis of Hirayama disease(1). Even in the study quoted by Dr. S. Kumar(2) the duration of illness at presentation ranged from 3- 64 months with 25% patients having duration of illness less than 12 months.

   Given the reported arrest of disease progression with the use of a cervical collar(2), it is imperative that the diagnosis is made as early as possible rather than waiting for a period of "at least 4-5yrs. of symptoms".
2. The patient reported had asymmetric