

Graves Disease in an Eight Year Old Boy of Down Syndrome

Down's syndrome is one of the most common chromosomal disorders in pediatric practice(1). The most common thyroid disorder with Down's syndrome is hypothyroidism(2)

An 8-year-old boy of Down's syndrome was referred to the Endocrine clinic with weight loss of three kilograms, irritability, change in behavior and restlessness. At presentation the child was irritable and restless, unable to sit still. He had up slanting eyes, epicanthic folds, high arched palate and protruding tongue. His height was 128 cm (90th percentile by the Indian Council Medical Research charts)(3). The weight was 20 Kg [50th percentile by ICMR charts]. He had a visible goiter, a bruit over the thyroid, exophthalmos and mental retardation. The mental age was 3.4 years. The serum Total Thyronine (T3) was 320 ng/dL (normal 55-180 ng/dL), Serum Thyroxin (T4) was 16 (µg/dL (normal 5.5-11(µg/dL). The Serum Thyrotropin (Serum TSH) was undetectable by third generation Immuno Radiometric assay. The radioiodine I¹³¹ uptakes at two hours was 76% and at twenty-four hours was 91.6%. Thyroid scan using Tc99 showed a diffuse uptake. The thyroid antimicrosomal antibody titers were not raised. He was treated initially with carbimazole 15 mg/day and propranolol 20 mg/day. There was symptomatic improvement after six weeks of treatment. The antithyroid drugs were continued for a period of eighteen months

with regular monitoring of the thyroid function. There was a gain in weight by 7 Kg from the pre treatment weight. The child responded with clinical and biochemical remission after 18 months of treatment with carbimazole. The child has maintained remission on subsequent follow up for six months.

Thyroid disorders are common in Down's syndrome. The anti thyroid auto antibodies are found in 13 to 34% of patients of Down's syndrome(2) Hypothyroidism due to autoimmune thyroiditis is the most common thyroid disorder. Hyperthyroidism is rare in the patients of Down's syndrome and the reported incidence of hyperthyroidism varies from 0.07- 2.5%. It rarely presents before eight years of age(4). The treatment is with a beta blocking agent such as propranolol, 1mg/kg/day in divided doses, and an antithyroid drug started at the same time. Carbimazole 0.5 to 1.0 mg/kg/day is given till the time remission is achieved(5). Our patient had Down's syndrome developed Graves disease that responded well with medical treatment. The rarity of this association especially in a male at eight years of age has prompted us to report this case. We recommend that thyroid function be followed closely in all the patients of Down's syndrome.

**Abhay I. Ahluwalia,
Shankar Narayan,**

Correspondence to:

Surg Commander Abhay I. Ahluwalia,

Classified Specialist,

Medicine & Endocrinology,

INHS Asvini,

Colaba, Mumbai 400 005, India.

E-mail: abhayahlu@hotmail.com

REFERENCES

1. Jain R, Thomas DC, Ragas R. Down syndrome: still a social stigma. *Am J Perinatol* 2002 19: 99-108.
2. Loudon MM, Day RE and Duke EM Thyroid dysfunction in Down's syndrome *Arch Dis Childh*, 1985; 60 : 1149-1151.
3. Technical Report Series No 18 ;Growth and Development of Indian Infants and Children: Indian Council of Medical Research Publication; 1989; pp. 61-136.
4. Karlsson B, Gustafsson J, Hedov G, Ivarsson SA, Anneren G. Thyroid dysfunction in Down's syndrome: Relation to age and thyroid autoimmunity. *Arch Dis Child* 1998 ; 79: 242-245.
5. Annotation Treatment of hyperthyroidism in young people *Arch Dis Child* 1998; 78: 207-209.

Role of IVIG in Preventing Exchange Transfusions in Rh Hemolytic Disease

Intravenous immunoglobulin (IVIG) treatment has been reported to decrease requirements for exchange transfusion, phototherapy, and to shorten hospitalization time for patients with Rh hemolytic disease of the newborn(1-3). It was shown that IVIG is also effective in prevention of repeated exchange transfusions when used after the first exchange transfusion(4). Here we report four cases of hemolytic disease in which exchange transfusion was indicated, but the patients were initially treated with IVIG.

Case 1 was a male infant. At birth, the infant did not show marked icterus but mild skin edema and hepatosplenomegaly were noted. The cord blood hemoglobin (Hb) level was 10.3 g/dL, and the total bilirubin level was 4.4 mg/dL. IVIG was administered. Four hours later, the patient's Hb and total bilirubin levels were 11.2 g/dL and 5.3 mg/dL, respectively.

Case 2 was a boy. On admission to our hospital at 26 hours after birth, the patient was icteric and his liver was palpable 4 cm below the costal margin. The serum Hb was

12 g/dL, the total bilirubin level was 19 mg/dL. He was given IVIG and phototherapy was initiated. Six hours later, the baby's Hb level was 13 g/dL and his total bilirubin had dropped to 14 g/dL.

Case 3 was a female. On admission to our hospital 10 hours after birth, the infant was icteric and both her liver and spleen were enlarged. The serum Hb level was 11 g/dL, total bilirubin was 10.5 mg/dL. The baby was given IVIG and started on phototherapy. Four hours later, her Hb level was 11.5 g/dL and her total bilirubin had fallen to 9 mg/dL.

Case 4 was a girl. Physical examination at birth revealed hepatosplenomegaly. She was given phototherapy in the first 24 hours of life. On admission to our hospital at 24 hours after delivery, the patient's serum Hb was 10 g/dL, and total bilirubin was 18 mg/dL. The baby was given IVIG. Six hours later, her Hb had risen to 12 g/dL, and her total bilirubin level was 16 mg/dL.

The four patients in this report developed hemolytic disease due to Rh-incompatibility. Exchange transfusion was indicated but was withheld, and treatment with 0.5 g/kg IVIG significantly reduced the rate of hemolysis in all cases. Only one of the four patients required subsequent red cell transfusions.