CASE REPORT: A HEALTHY DAUGHTER BORN TO A CRETIN.

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Thyroxine replacement therapy can cure congenital hypothyroidism (cretinism) if instituted early in life. We like to report a case of healthy daughter born to a congenitally hypothyroid woman considering its rarity.

CASE REPORT

A girl aged 11 months was put on thyroxine therapy in 1971 by the renowned pediatrician Prof. Dr. M. R. Khan who diagnosed her to be congenitally hypothyroid. The initial dose was 12.5 microgram (mcg) daily and gradually it was increased to 150 mcg/day in adult life. She was highly educated (Master of Science) and was married on 18 August/2000 and got pregnant in 2001 when her thyroxine dose was increased further upto 200 mcg/day. She gave birth to a healthy daughter on 03 January/2002 by Caesarean section due to transverse lie. Cord blood hormones were assayed: T_3=0.76 nmol/L (normal range 0.8-3.16), T_4=113 nmol/L (normal range 64-175), TSH=6 mIU/L (normal range 0.4-5). However, she was euthyroid clinically on 13.03.2002.

DISCUSSION

Permanent primary congenital hypothyroidism affects about one newborn in 3500. Eighty to ninety percent of the cases are due to developmental defects of the thyroid gland (thyroid dysgenesis), such as arrested migration of the embryonic thyroid (ectopic thyroid) or a complete absence of thyroid tissue (athyreosis). Most cases of thyroid dysgenesis are sporadic and result from as yet unknown mechanisms. The remaining 10-20% have functional defects in one of the steps involved in thyroid-hormone biosynthesis (thyroid dyshormonogenesis)-defects transmitted by an autosomal recessive mode of ingeritance.[1] The pathogenesis of thyroid dysgenesis is not known.[2] Healthy baby born to a hypothyroid mother is a rare phenomenon, however, early treatment and regular monitoring of hormone levels may lead to an absolutely normal life. In 1989, a starting dose of 10-15 mcg./kg per day of thyroxine was proposed and has been widely used since.[3] The upper range of normal values for plasma free thyroxine in normal infants is much higher than that for older children or adults.[4] Premature fusion of the fontanelles, a recognised complication of perinatal hyperthyroidism (such as seen in children born to mothers with Graves' disease), had never been reported in infants with congenital hypothyroidism treated with 10-15 mcg/kg per day of levothyroxine.[1] During pregnancy and estrogen therapy the need for thyroxine is increased.[5] Screening of neonates for congenital hypothyroidism is being done in many conutries,[6] but screening of pregnant women for hypothyroidism is not yet universal.[7] Haddow et al and Utiger encouraged adequate iodine intake and it should be increased during pregnancy.[8,9]

REFERENCES

1. Van Vliet G. Treatment of congenital hypothyroidism. Lancet 2001;358: 86-87.

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2. Castanet M, Lyonnet S, Bonaiti-Pellie C et al. Familial forms of thyroid dysgenesis among infants with congenital hypothyroidism. N Engl J Med. 2000; 343: 441-442.
3. Fisher DA, Foley BL. Early treatment of congenital hypothyroidism. Pediatrics 1989; 83: 785-89.
4. Zurakowski D, Di Canzio J, Majzoub JA. Pediatric reference intervals for serum thyroxine, triiodothyronine, thyrotropin, and free thyroxine. Clin Chem 1999; 45: 1087-91.
5. Arafah BM. Increased need for thyroxine in women with hypothyroidism during estrogen therapy. N Engl J Med 2001; 344: 1743-9.
6. Taher MA. Follow-ups of congenital hypothyroidism. ASEAN J Radiol 2001; VII(ii): 121-3.
7. Pop VJ, Van Baar AL, Vulsma T. Should all pregnant women be screened for hypothyroidism? Lancet 1999; 354; 1224-5.
8. Haddow JE, Palomaki GE, Allan WC, et al. Maternal thyroid deficiency during pregnancy and subsequent neuropsychological development of the child. N Engl J Med. 1999; 342: 549-55.
9. Utiger RD. Maternal hypothyroidism and fetal development. N Engl J Med 1999; 341: 601-02.