
COMMUNICATIONS:**3. CASE REPORT: TWO HEALTHY DAUGHTERS
BORN TO A CRETIN****Dr. Muhammad Abu Taher****INTRODUCTION**

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 Professor 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 she took 175 mcg/day of thyroxine and her daughter was born on 03 January/2002 by Cesarean 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. On 9 Feb. 2004, she had another sister born on 22 Dec. 2003 whose TSH is 9.5 mIU/L and she is also euthyroid clinically and all three (mother and both daughters) are on long-term follow-ups.

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 dysmorphogenesis)-defects transmitted by an autosomal recessive mode of inheritance.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 proposed³ and has been widely used since then, however, a recent systematic review does not support clinical recommendation of high or standard starting doses of levothyroxine.⁴ The upper range of normal values for plasma free thyroxine in normal infants is much higher than that for older children or adults.⁵ 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.⁶ Screening of neonates for congenital hypothyroidism is being done in many countries, but screening of pregnant woman for hypothyroidism is not yet universal.⁸ Haddow et al. and Utiger encouraged adequate iodine intake and it should be increased during pregnancy.^{9,10} In North America, screening programs usually initially measure T₄ and for those with low T₄ levels, TSH is then measured on the initial sample. In Europe and Japan, screening programs often use TSH elevation as the initial screening test, but this approach does not detect pituitary insufficiency. Transient neonatal hypothyroidism may also occur in infants of mothers with autoimmune thyroid disease and is due to maternal antibodies, particularly TSH-binding inhibitory antibody. A thyroid with normal morphology and normal or decreased uptake may be seen with transient hypothyroidism, and this can be evaluated by discontinuing thyroid replacement. Such a trial is usually not performed until 3 years of age so that the child is not subjected to thyroid insufficiency while the thyroid is needed for neurologic development.¹¹

REFERENCES

1. Van Vliet G. Treatment of congenital hypothyroidism. *Lancet* 2001; 358: 86-87.
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;343:441-44
3. Fisher DA, Foley BL, Early treatment of congenital hypothyroidism. *Pediatrics* 1989; 83:785-89.
4. Hrytstuck I, Gilbert R, Logan S, Pindoria S, Brook CGD. Starting dose of levothyroxine for the treatment of congenital hypothyroidism. A systematic review. *Arch Pediatr Adolesc Med* 2002; 156:485-491.
5. Zurakowski D, Di Canzio J, Majzoub JA. Pediatric reference intervals for serum thyroxine, triiodothyronine, thyrotropin, and free thyroxine. *Clin Chem*. 1999; 45:1087-91.
6. Arafah BM. Increased need for thyroxine in women with hypothyroidism during estrogen therapy. *N Engl J Med* 2001; 344:1743-9.
7. Taher MA. Follow-ups of congenital hypothyroidism. *ASEAN J Radiol* 2001; VII (ii): 121-3.
8. Pop VJ, Van Baar AL, Vulsma T. Should all pregnant women be screened for hypothyroidism? *Lancet* 1999; 354:1224-5.
9. Haddow JE, Plaomaki GE, Allan WC, et al. Maternal thyroid deficiency during pregnancy and subsequent neuropsychological development of the child. *N Eng J Med* 1999; 342: 549-55.
10. Utiger RD. Maternal hypothyroidism and fetal development. *N Engl J Med* 1999; 341:601-02.
11. Shore RM. *Pediatrics*. In Wilson MA (ed.): *Text book of Nuclear Medicine*, 1998, Lippincott-Raven, Philadelphia pp. 259-277.