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Research Article

Iodine Deficiency Cretinism in Tuva Republic, Siberia

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Abstract

Introduction: Iodine deficiency is the most common cause of mental retardation that can be prevented. The most serious consequence of iodine deficiency is cretinism.

The Republic of Tuva is one of 89 administrative territories of the Russian Federation. Tuva lies in the south of Siberia and bordering Mongolia. In 1997 we first discovered the pocket of severe iodine deficiency in the Republic of Tuva and found cases of endemic cretinism.

The Aim: To study the features of iodine deficiency cretinism in the Republic of Tuva.

Materials and Methods: We surveyed 287 children between the ages of 1.5 and 15 in the western districts of Tuva. A neurological examination was carried out; audiometry, ultrasound examination of hearing; psychometric assessment of mental development through verbal and non-verbal tests.

Results: According to our research, the prevalence of iodine deficiency cretinism in Tuva was 3.5%. Various forms of endemic cretinism have been revealed: mixedematous, neurological and mixed. Mixed cretinism prevailed (77.6%) and characterized signs of severe hypothyroidism, mental and growth retardation, emotional-will disorders. Patients with neurological cretinism were deafness in 75%, with serious mental deficit (idiocy or imbecility) combined with squint and characteristic spastic lesions of the limbs.

Conclusions: Severe iodine deficiency in Tuva Republic has a negative impact on the health and mental development of the population and requires constant adequate iodine prevention and monitoring of iodine deficiency.

Keywords: Endemic Cretinism; Iodine Deficiency Disease; Congenital Hypothyroidism

Abbreviations

TSH: Thyroid Stimulating Hormone; T4: Thyroxine.

Introduction

The iodine deficiency cretinism was not reported in Russia after the early 1950's, when the national control program began the large scale distribution of iodized salt and iodine tablets to populations of 2 iodine goiter regions" as defined by the Ministry of Health. Tuva is the first district of Russia where new cases of iodine deficiency cretinism are been discovered.

The Republic of Tuva is one of 89 administrative territories of the Russian Federation. It lies in the geographical center of Asia bordering Mongolia. Of its 324,421 people, 80.9% are indigenous Tuvinians. When I came to Tuva for the first time in 1997, the village Chaa-Hol was so quiet, were no activities, everyone looked lethargic and gave the impression of been lazy. Then we found out that it was related with hypothyroidism of this population (26%).

Beginning in 1997, several surveys by doctors from the Institute for Medical Studies of the North (Krasnoyarsk) revealed severe iodine deficiency in the west of the republic, characterized by a high endemic goiter prevalence, congenital and non-congenital hypothyroidism, endemic cretinism (3.5% prevalence), a very low urinary iodine (16 mcg/l), high serum thyroglobulin (median was 86 ng/ml) and a high transient neonatal hypothyroidism prevalence (15.0 - 34.2%) on screening [5-9]

The condition of endemic cretinism is defined by three major features [1]:

- Epidemiology: It is associated with endemic goiter and severe iodine deficiency.
    - Clinical manifestations: These comprise mental deficiency together with either:
    - A predominant neurological syndrome consisting of defects of hearing and speech, and with characteristic disorders of stance and gait of varying degree: or
    - Predominant hypothyroidism and stunted growth.
    - Although in some regions one of the two types may predominate, in other areas a mixture of the two syndrome will occur.
- Prevention: In areas where adequate correction of iodine deficiency has been achieved, endemic cretinism has been prevented.

The three characteristic features of neurological endemic cretinism are extremely severe mental deficiency together with squint, deafmutism and motor spasticity with disorders of the arms and legs of a characteristic nature. Dr. G. Robert DeLong [2] suggests that the neuropathological basis of the clinical picture includes underdevelopment of the cochlea for deafness; maldevelopment of the cerebral neocortex for mental retardation; and maldevelopment of the corpus striatum (especially putamen and globus pallidus) for the motor disorder.

The typical myxedematous cretin has a less severe degree of mental retardation than the neurological cretin, but has all the features of severe hypothyroidism present since early life, as in untreated sporadic congenital hypothyroidism: severe growth retardation, incomplete maturation of the facial features including the naso-orbital configuration, atrophy of the mandibles, puffy features, myxedematous, thickened and dry skin, dry and decreased hair, eyelashes and eyebrows and much delayed sexual maturation. Contrasting with the general population and with neurological cretinism, goiter is usually absent and the thyroid is often not palpable, suggesting thyroid atrophy [3,4].

Materials and Methods

We surveyed 287 children between the ages of 1.5 and 15 in the western districts of Tuva. A neurological examination was carried out; audiometry, ultrasound examination of hearing; psychometric assessment of mental development through verbal and non-verbal tests.

Results and Discussion

According to our research, the prevalence of iodine deficiency cretinism in Tuva was 3.5%. We revealed various forms of endemic cretinism: mixedematous and neurological. Mixedematous cretinism prevailed (77.6%) and characterized typical clinical picture of congenital hypothyroidism: mental and growth retardation, emotional-will disorders. Although more than 15% of neonatal blood samples had TSH levels >20 mU/L, most of these cases were transient and not treated as permanently hypothyroidism.

Neurological cretinism has the three characteristic features of: extremely severe mental deficiency together with squint, deafmutism and motor spasticity with disorders of the arms and legs of a characteristic nature. Neurological cretins were deaf, with serious mental deficit (idiocy or imbecility) combined with squint and characteristic spastic lesions of the limbs (featured by tight hips and thighs, increased knee and adductor reflexes, slow facial movements and smile, flexed gait, shuffling; motor rigidity, spasticity with increased reflexes and ankle clonus), speech disorders and mental retardation.

In the Chaa-Hol village we found 28 cases of iodine deficiency cretinism. Some cases are here:

- Female, 11 years: short stature, poor memory, TSH 480 mU/L (normal 0.4-4.0 mU/L), bone age 3 years. Diagnosis: congenital hypothyroidism, mixedematous cretinism.
- Male, 3.5 years, congenital hypothyroidism diagnosed at 1.8 years, TSH 801 mU/L (normal 0.4-4.0 mU/L), could not walk or sit at age 2; began walking at age 2.1. Diagnosis: congenital hypothyroidism, mixedematous cretinism.
- Family with congenital hypothyroidism, mixedematous cretinism (mother: nodular goiter, hypothyroidism):
  - 5 years daughter, congenital hypothyroidism diagnosed at 2.5 years; T4 – zero; TSH 218 mU/L.
  - 3 years son, congenital hypothyroidism diagnosed at 10 months; TSH 303 mU/L.

These dramatic findings of cretinism confirmed severe iodine deficiency in Tuva and called for urgent implementation of control programs. In 1998, the Tuva Government and Ministry of Health, in collaboration with scientists from the Institute for Medical Studies of the North, developed the program for eliminating of iodine deficiency in the republic, which included iodized salt consumption for the entire population and iodine pills for high risk groups. Iodine deficiency was eliminated in the Tuva Republic in 2000. KIWANIS International built salt iodination factory in capital Kyzyl in 2002 [5-9].

Figure 1: Myxedematous endemic cretinism in Tuva (female, 40 years).

Figure 2: Exploring iodine deficiency cretinism in Tuva Republic (in center Dr. G.R. DeLong and Dr. I.V. Osokina).

Conclusion

Severe iodine deficiency in Tuva Republic has a negative impact on the health and mental development of the population and requires constant adequate iodine prevention and monitoring of iodine deficiency.

Bibliography

1. Stanbury JB. "The iodine trial. Exploring iodine deficiency and its prevention around the world". Oxford University Press (2008): 1-202.

2. DeLong RG. "Neurological involvement in iodine deficiency disorders". In : Hetzel BS, Dunn JT, Stanbury JB (eds): The prevention and control of Iodine Deficiency Disorders. Amsterdam, Elsevier (1987): 49.

3. Chan S and Kilby MD. "Thyroid hormone and central nervous system development". *Journal of Endocrinology* 165 (2000): 1.

4. Osokina IV. "Iodine Deficiency in Central Siberia". Exploring, prevention and monitoring. Palmarium academic publishing, Germany. (2013): 1-234.

5. Osokina IV and Manchouk VT. "Severe Iodine Deficiency in Tuva Republik". *IDD Newsletter* 14 (1998): 59-60.

6. Osokina I., *et al*. "The current status of iodine deficiency disorders in Tyva Republic, Russian Federation". *IDD Newsletter* 17 (2001): 58-60.

7. Osokina IV. "Epidemiological and Immunogenetic Peculiarities of Type 1 Diabetes and Iodine Deficiency Disorders in Central Siberia". Author's abstract of PhD thesis. Moscow (2002): 1-39.

8. Osokina IV and Manchouk VT. "Iodine deficiency disorders in Siberia". Novosibirsk, Science (2012): 1-153.

9. Osokina Irina V. "Iodine deficiency in Central Siberia, Russia". *Acta Scientific Nutritional Health* 3 (2019): 127-129.

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