The Turkish Journal of Pediatrics 1998; 40: 431-435

COMPLETE ATRIOVENTRICULAR HEART BLOCK IN CONGENITAL HYPOTHYROIDISM*

*Rana Olguntürk MD**, F. Sedef Tunaoğlu MD***, Deniz Oğuz MD****
*Peyami Cinaz MD*****, Aysun Bideci MD******

SUMMARY: Olguntürk R, Tunaoğlu FS, Oğuz D, Cinaz P, Bideci A. (Departments of Pediatric Cardiology and Endocrinology, Gazi University Faculty of Medicine, Ankara, Turkey). Complete antrioventricular heart block in congenital hypothyroidism. Turk J Pediatr 1998; 40: 431-435.

Severe hypothyroidism in children is known to produce cardiac abnormalities such as asymmetric thickening or hypertrophy of the interventricular septum, smaller internal dimensions of the left ventricle, a smaller left ventricular outflow tract, and less systolic septal excursion. In this report, we present a 1.5-year-old boy who was admitted to our hospital because of growth retardation. According to the clinical and laboratory findings, congenital hypothyroidism, dilated cardiomyopathy (DCMP), atrioventricular complete heart block and secundum type atrial septal defect were diagnosed. *Key words: AV block, hypothyroidism, congenital.*

Severe hypothyroidism has long been known to produce abnormalities of cardiac structure and function[1]. Morphologic abnormalities such as asymmetric thickening or hypertrophy of the interventricular septum, smaller internal dimensions of the left ventricle, a smaller left ventricular outflow tract, and less systolic septal excursion have been reported in hypothyroid adults and children[2-4]. However, there is much less reported and contradictory data about disturbances of cardiac function of infants with hypothyroidism[5, 6]. In this report, we present a 1.5-year-old boy with congenital hypothyroidism who had dilated cardiomyopathy (DCMP), atrioventricular complete heart block and secundum type atrial septal defect.

Case Report

A 1.5-year-old boy was admitted to Gazi University Hospital because of growth retardation. His mother was a healthy woman. His parents were consanguineous and had a five-year-old healthy son. His physical examination revealed weight

* From the Departments of Pediatric Cardiology and Endocrinology, Gazi University Faculty of Medicine, Ankara.

** Professor of Pediatrics and Pediatric Cardiologist, Gazi University Faculty of Medicine.

*** Associate Professor of Pediatrics and Pediatric Cardiologist, Gazi University Faculty of Medicine.

**** Pediatric Cardiologist, Gazi University Faculty of Medicine.

***** Associate Professor of Pediatrics and Pediatric Endocrinologist, Gazi University Faculty of Medicine.

****** Pediatrician, Gazi University Faculty of Medicine.

6100 g (< 3rd percentile), length 70 cm (< 3rd percentile), coarse facial appearance, large tongue, skin mottling, hypotonia, umbilical hernia, and pes equinovarus deformity. He was inactive, unsmiling and reluctant to contact, and was crying hoarsely. His skin was smooth and his hair was not coarse. His hematologic values were: hemoglobin 10.6 g/dl, ferritin 5.14 µg/L and hypochromia in blood smear. He had iron deficiency anemia. His chest x-ray showed cardiomegaly, and ECG showed atrioventricular complete heart block, ventricular rate 44/min and low voltage (Fig. 1a). Echocardiographic and angiocardiographic examinations displayed secundum type atrial septal defect, (QP/QS = 1.42) tricuspid insufficiency (1°) and dilated cardiomyopathy, but no pericardial effusion. His echocardiographic measurements are shown in Table I. His bone age was three months according to his extremity x-rays. Thyroid function tests were performed by RIA and TSH was performed by IRMA methods. The serum thyroxine (T4) was 1.74 µg-dl (N: 4.5-1.25 µg-dl), T3 0.44 ng/dl (N: 0.52-1.75 ng/dl), and TSH 50 mIU/ml (N: < 5.5 mIU/ml).

Based on his physical and laboratory examinations, he was diagnosed as having hypothyroidism and treated with gradually increasing doses of thyroxine starting from 0.025 mg daily, up to 0.100 mg daily.

After one month of thyroxine therapy he was smiling, and he began to sit and play. His echocardiographic measurements improved (Table I). His heart rate was 75/min, and he still had atrioventricular complete heart block (Fig. 1b). Thyroid function test results were: T4 value 10.62 µg/dl, T3 value 0.68 ng/dl, free T4 1.65 ng/dl (N: 0.73-1.95 ng/dl), free T3 2.67 pg/ml (N: 2.2-4.7 pg/ml), and TSH 6.69 mIU/ml.

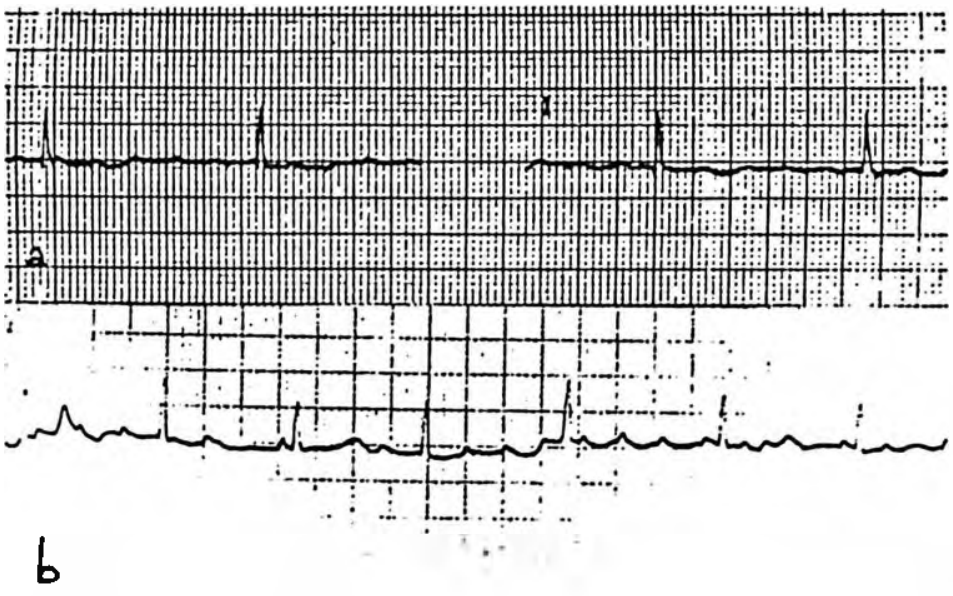

Fig. 1: a) ECG, pretreatment. b) ECG, one month after the start of therapy.

Table I: Echocardiographic Measurements

Values	Before Therapy	First Month of Therapy	Normal Values (Min-Max)
LVDd (cm	3.90	3.50	2.20-2.70
LVDs (cm)	3.00	2.20	1.30-1.90
LVFS (%)	23	37	0.28-0.44
LVVd (mm^3)	9.74	10.81	
LVVs (mm^3)	6.53	4.24	
LVEF (%)	33	61	64-83
PWd (cm)	0.30	0.50	0.4-0.55
VSd (cm)	0.30	0.50	0.4-0.5
PWs (cm)	0.60	0.80	
VSs (cm)	0.70	0.80	
PEP (msc)	56.0	64.0	$< 131 \pm 13$
ET (msc)	240	267	$< 415 \pm 11$
PEP/ET (%)	23	24	0.30-0.39

LVDd : left ventricular dimension in diastole.
LVDs : left ventricular dimension in systole.
LVFS : left ventricular shortening fraction.
LVVd : left ventricular diastolic volume.
LVVs : left ventricular systolic volume.
LVEF : left ventricular ejection fraction.
PWd : left ventricular posterior wall dimension in diastole.
VSd : ventricular septum dimension in diastole.
PWs : left ventricular posterior wall in systole.
VSs : ventricular septum dimension in systole.
PEP : pre-ejection period.
ET : ejection time.

Discussion

Our patilent's clinical and laboratory findings revealed the diagnosis of dilated cardiomyopathy, and his systolic time intervals were also abnormal. The latter findings have been previously reported in children and adults with hypothyroidism[6,7]. Though there were no reports on dilated cardiomyopathy in children with hypothyroidism, Ladenson et al.[8] had reported a young man with hypothyroidism and DCMP who showed marked improvement in left ventricular ejection fraction (LVEF), (LVIDd), and cardiac index. They suggested that the alteration in gene expression had caused the dilated myopathic heart. In our patient, the investigation with gene expression was not performed; it can be suggested that the long duration of the hypothyroidic state was the cause of DCMP.

The complete atrioventricular block was the other unusual finding in our patient. Alterations in heart rate, first degree heart block and bundle branch block are the conduction disorders so far defined in hypothyroidism[9,10]. recently accumulating data indicates that atrioventricular block is a complication of amiodarone-induced hypothyroidism[11]. Electrophysiological studies suggested the presence of dual atrioventricular nodal pathways in a patient with hypothyroidism[12]. We reached no conclusion about AV nodal pathways, as the electrophysiological study was not performed on our patient. Syed[13] reported

an infant with congenital hypothyroidism and atrioventricular block whose conduction disturbance persisted despite therapy. This is the only case with AV block we found reported in the literature. Baysal et al.[14] reported that echocardiographic pathologic findings of children with hypothyroidism returned to normal with thyroxine therapy. In our patient, we observed significant improvements in echocardiographic measurements during thyroxine therapy, but atrioventricular block was continuing. It has been suggested that cardiac function disturbances in adults with hypothyroidism are exaggerated when associated with other pathologies, like atherosclerosis[15]. Our patient had secundum type atrial septal defect, but it is known that there is no association between atrial septal defect and complete heart block.

In our patient, the improvement of cardiac function with thyroxine therapy indicates that hypothyroidism was responsible for his dilated cardiomyopathy. His conduction disturbance may be congenital, or due to his prolonged hypothyroidic state. We can also assume that the effects of hypothyroidism worsened the conduction disturbances as is the case with adults[16].

REFERENCES

1. Crowley WF Jr, Ridgway EC, Bough EW, et al. Noninvasive evaluation of cardiac function in hypothyroidism. Response to gradual thyroxine replacement. N Engl J Med 1977; 296: 1-6.
2. Farooki ZQ, Hoffman WH, Perry BL, Green EW. Myocardial dysfunction in hypothyroid children. Am J Dis Child 1983; 137: 65-68.
3. Balducci G, Acquafredda A, Amendola F, Natuzzi M, Laforgia N, Cavallo L. Cardiac function in congenital hypothyroidism: impairment and response to L-T4 therapy. Pediatr Cardiol 1991; 12: 28-32.
4. Santos AD, Miller RP, Mathew P, Wallace WA, Cave WT Jr, Hinojosa L. Echocardiographic characterization of the reversible cardiomyopathy of hypothyroidism. Am J Med 1980; 68: 675-682.
5. Fouron JC, Bourgin JH, Letarte J, Dussault JH, Ducharme G, Dawignon A. Cardiac dimensions and myocardial function of infants with congenital hypothyroidism. An echocardiographic study. Br Heart J 1982; 47: 584-587.
6. Hayford JJ, Schieken RM, Thompson RG. Cardiac function in primary hypothyroidism. Am J Dis Child 1980; 134: 556-559.
7. Burckhardt D, Staub JT, Kraenzlin M, Raeder E, Engel U, Cloppenburg P. The systolic time intervals in thyroid dysfunction. Am Heart J 1978; 95: 187-196.
8. Ladenson PW, Sherman SI, Baughman KL, Ray PE, Feldman AM. Reversible alterations in myocardial gene expression in a young man with dilated cardiomyopathy and hypothyroidism. Proc Natl Acad Sci 1992; 89: 5251-5255.
9. Lardoux H, Cenac A, Perlemuter L, Bernheim R, Hazard J. Troubles de conduction intracardique et hypothroidie de l'adulte. Etude systematique de 42 cs. Nouv Presse Med 1975; 4: 1859-1862.
10. Hansen JE. Paroxysmal ventricular tachycardia associated with myxedema. A case report. Am Heart J 1961; 61: 692.
11. Mangiardi L, Gaita F, Brun S, Presbitero P, Nademanee K, Singh BN. Atrioventricular block complicating amiodarone-induced hypothyroidism in a patient with pre-excitation and rate-dependent bilateral bundle branch block. J Am Coll Cardiol 1986; 7: 180-184.

12. Denes P, Wyndham CR, Wu D, Rosen KM. "Supernormal conduction" of premature impulse utilizing the fast pathway in a patient with dual atrioventricular nodal pathways. Circulation 1975; 51: 811-814.

13. Syed AA. Congenital heart block and hypothyroidism. Arch Dis Child 1978; 53: 256-258.

14. Baysal K, Özkutlu S, Saraçlar M, Binginturan N, Özer S. Echocardiographic evaluation of cardiac changes in children with hypothyroidism. Çocuk Sağlığı ve Hastalıkları Dergisi 1985; 28: 293-301.

15. Tajiri J, Morita M, higashi K, Fujii H, Nakamura N, Sato T. The cause of low voltage QRS complex in primary hypothyroidism. Pericardial effusion or thyroid hormone deficiency? Jpn Heart J 1985; 26: 539-547.

16. Singh JB, Starobin OE, Guerrant RL, et al. Reversible atrioventricular block in myxedema. Chest 1973; 63: 582-585.