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Hereditary Nephritis and the Heart

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Eleven cases of hereditary nephritis were studied for cardiac abnormalities by means of ECG and BCG. With the exception of two cases no significant abnormalities were demonstrable, which indicates that this genetically transmitted process generally leaves the heart unaffected. It is thus to be expected that intermittent haemodialysis, as far as the heart is concerned, will be well tolerated by these patients.

To the best of our knowledge the state of the heart in hereditary nephritis has received little attention thus far. The question whether this genetically determined process affecting the kidney and the auditory organ [1, 2, 3, 4, 5] also involves the heart has still remained unanswered.

Material and method

Eleven patients with hereditary nephritis were studied for cardiac abnormalities by means of ECG and BCG. There were 7 males, and 4 females, age ranging between 13 and 46 years. The mode of inheritance could be traced in the case of five families (Figs 1 to 5). It is to be noted that in family No. 5 the parents were healthy and had an only male child. In this case Alport's syndrome was found in association with tapetoretinal degeneration and complete atrioventricular block.

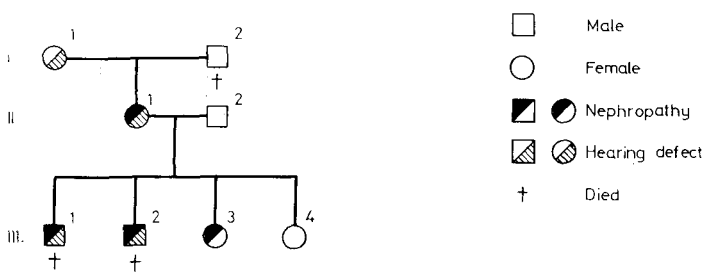

Fig. 1. Mode of inheritance in Family 1

The 12-lead ECG was recorded with an Elema Mingograph Type 42 and with a Biocomb-5 apparatus. For ballistocardiography a Bodrogi direct acceleration apparatus and a Schwarczer–Klensch ultra-low frequency, critically controlled

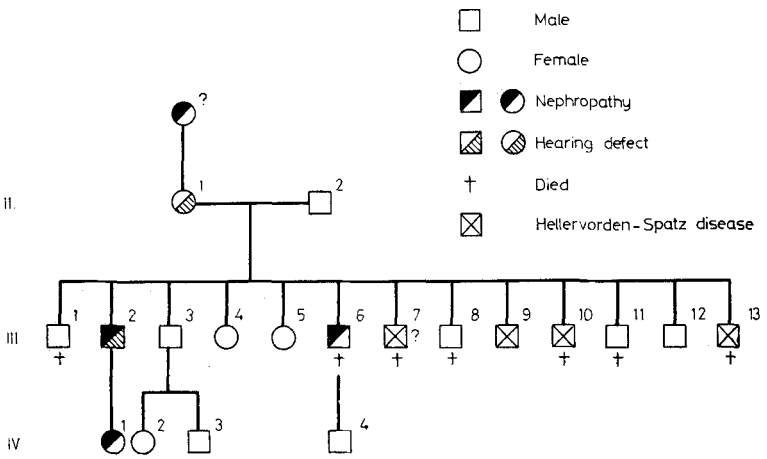

Fig. 2. Mode of inheritance in Family 2

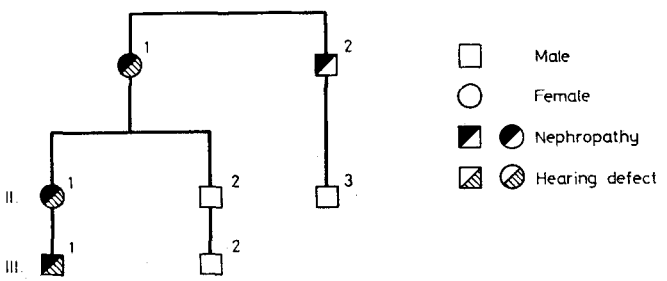

Fig. 3. Mode of inheritance in Family 3

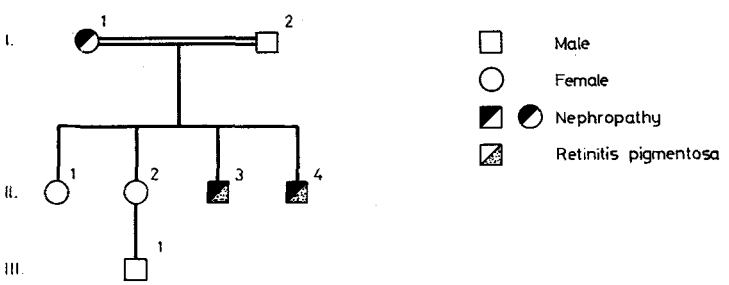

Fig. 4. Mode of inheritance in Family 4

BCG table were used. ECG, PCG, carotid-pressure or finger-pulse tracing served as reference curves. Our instruments registered the direct acceleration and the indirect displacement curves, respectively. The two curves basically represent the mechanical function of the heart in two different ways.

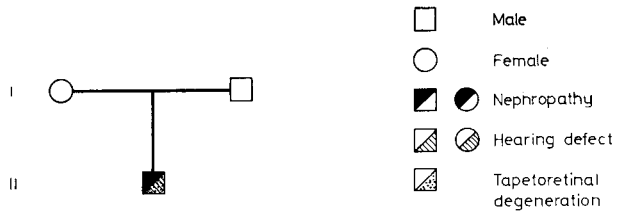

Fig. 5. Mode of inheritance in Family 5

Results

The ECG and BCG abnormalities of maximal severity demonstrable on repeated tracings are compiled in Tables 1 and 2.

As it can be seen, all patients were in sinus rhythm, with the exception of case 11 who had complete AV-block (Fig. 6). Regular sinus rhythm returned from time to time but the depolarization time remained 0.12 sec as before. PQ was prolonged in one patient (Case 7). QT referred to heart rate was prolonged in Cases 3 and 11. In the frontal plane, the mean P-vector was shifted to the right in 4 cases. It was vertical in 3 cases and showed a definite shift to the right in one

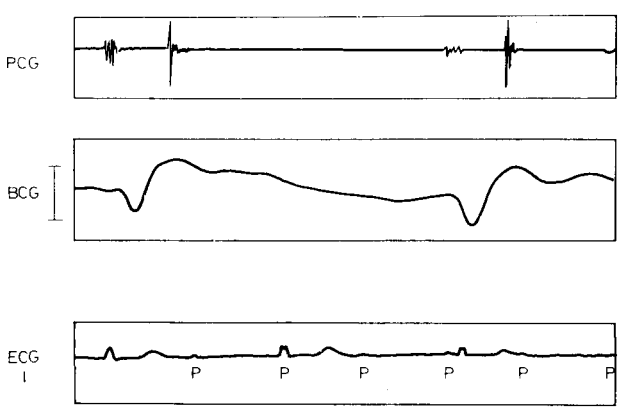

Fig. 6. P. Gy., 21-year-old male (Case 11). Diagnosis: Alport's syndrome. Tapetoretinal degeneration. ECG: complete AV-block. Indirect displacement BCG: normal. Standardization: 1 mV: 100 μ displacement. Paper speed: 50 mm/sec

Table 1

ECG data of 11 patients with

No.	Sex	Age (years)	Family	Rhythm	PQ	QRS	QT	Frontal projection	
					(sec)			P	R
1	F	46	1 II/1	Sinus	0.16	0.08	0.32	Steep	Mid-pos.
2	M	20	1 III/1	Sinus	0.16	0.06	0.38	Mid-pos.	Mid-pos.
3	M	20	1 III/2	Sinus	0.16	0.08	0.44	Steep	Mid-pos.
4	F	17	1 III/3	Sinus	0.16	0.08	0.28	Mid-pos.	Mid-pos.
5	M	37	2 III/2	Sinus	0.14	0.08	0.34	Mid-pos.	Mid-pos.
6	M	26	2 III/6	Sinus	0.13	0.08	0.32	Mid-pos.	Mid-pos.
7	F	13	2 IV/1	Sinus	0.22	0.06	0.38	Steep	Mid-pos.
8	F	27	3 II/1	Sinus	0.14	0.08	0.32	Mid-pos.	Mid-pos.
9	M	21	4 II/3	Sinus	0.18	0.08	0.36	Mid-pos.	Mid-pos.
10	M	18	4 II/4	Sinus	0.12	0.08	0.32	Right	Right
11	M	21	5 II/1	3° AV block	—	0.12	0.50	Mid-pos.	Left

M = male
F = female

case. Direction of the R-vector was normal in all but 2 cases. ST-depression or elevation was found sporadically. Flat T-waves were more common. Tall, peaked T-waves were also encountered. Inverted T-waves were found in one case only. In the course of treatment the flat T-waves became tall and peaked (Fig. 7).

Changes of major severity were thus confined to Case 11. We have, however, no satisfactory explanation for the aetiology of the complete AV-block in this case. It may have been acquired, since the ECG had been found normal one year earlier, but it might represent a particular form of Alport's syndrome since the antecedents lacked any indication of earlier carditis.

The abnormalities revealed by the acceleration BCG corresponded at the worst to grade I of the Brown classification, regarded by us as irrelevant. In 6 cases the changes were too discrete to be classified into the Brown-scheme or, indeed, to be regarded as abnormalities at all. These included high amplitudes, tall L-waves, slurred H- and J-waves.

Examined by the displacement BCG, the amplitude of the fast ejection phase, i.e. of segment I"–J" which is most representative of the force of ventricular contraction, was decreased in Case 9. The abnormality proved, however, reversible: it disappeared as a result of intermittent haemodialysis (Fig. 7). In sum, appreciable BCG abnormalities were confined to one case.

Three of our patients (Cases 2, 3 and 6) died of renal failure. Apart from the anaemia and a slight fatty degeneration the myocardium revealed no particular abnormality.

hereditary nephritis

of mean vectors	ST		T			
T	Depression	Elevation	Flat	Biphasic	Inverted	Tall, peaked
Mid-pos.	—	—	—	—	—	—
Mid-pos.	II, V4	—	aVL I	II, V6	—	—
Mid-pos.	—	—	V6	—	—	V3, V4
Mid-pos.	—	—	—	+	—	—
Mid-pos.	—	—	—	—	—	—
Mid-pos.	—	—	aVL, I	—	—	—
Left	—	—	—	—	—	—
Mid-pos.	—	—	I, II, V3	—	—	—
Mid-pos.	II, V4, V5, V6	—	in all leads	—	—	—
Left	—	—	V6	—	—	—
Right	aVL I, V5–V6	aVF III, VL	—	—	aVL I, V2–6	

Table 2

BCG data of 11 patients with hereditary nephritis

No.	Sex	Age (years)	Family	Ballistocardiogram		
				Direct acceleration	Indirect displacement	
					I"–J" ampl.	I"–M" ampl.
1	F	46	1 II/1	Tall H	—	—
2	M	20	1 III/1	High amplitudes	—	—
3	M	20	1 III/2	Brown I	—	—
4	F	17	1 III/3	Slurred H and J	—	—
5	M	37	2 III/2	Tall L	—	—
6	M	26	2 III/6	High amplitudes	—	—
7	F	13	2 IV/1	Normal	Normal	Normal
8	F	27	3 II/1	Brown I	—	—
9	M	21	4 II/3	—	Reduced	Reduced
10	M	18	4 II/4	Tall diastolic deflections	Normal	Reduced
11	M	21	5 II/1	—	Normal	Normal

M = male
F = female

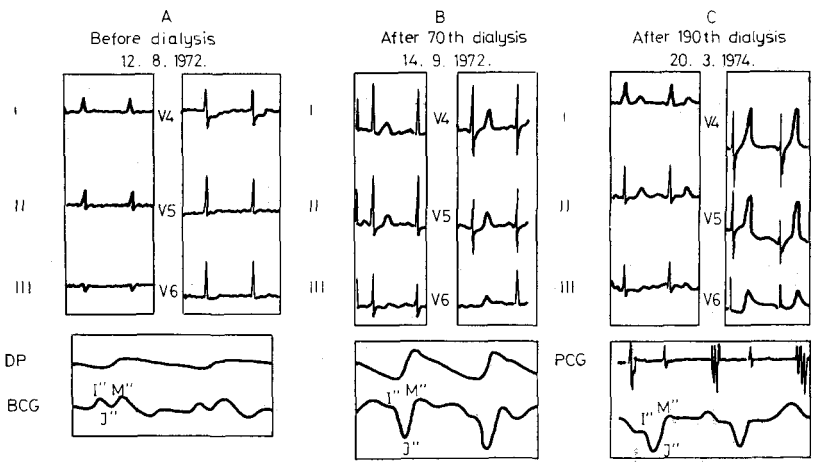

Fig. 7. ECG and indirect displacement BCG in the course of intermittent haemodialysis. V. F., 21-year-old male (Case 9). Diagnosis: Alport's syndrome. Renal failure. (A) Blood pressure: 170/100 mm Hg. BUN: 131 mg/100 ml; serum creatinine: 9.2 mg/100 ml; serum K: 3.6 mEq/l. ECG: sinus rhythm, no R-axis deviation, flat T-deflections. BCG: abnormal, the amplitudes of J" and M" being markedly reduced. (B) Blood pressure: 130/80 mm Hg. BUN: 135 mg/100 ml; serum creatinine: 16 mg/100 ml; serum K: 5.9 mEq/l. ECG: sinus rhythm, no R-axis deviation, high voltage, tall, peaked T-deflections. BCG: J" normal, M" low. (C) Blood pressure: 150/80 mm Hg. BUN: 151 mg/100 ml; serum creatinine: 17.4 mg/100 ml; serum K: 5.2 mEq/l. ECG: sinus rhythm, no R-axis deviation. The T-amplitudes in the chest leads have increased. BCG: normal J", low M". Standardization of the indirect displacement BCG: 1 mV: 100 μ displacement. Paper speed: 50 mm/sec

Discussion

From the present study it emerges that in hereditary nephritis neither the electric nor the mechanic functions of the heart are significantly affected. Possible explanations are the following:

1. The patients were fairly young. In our age-matched cases of chronic glomerulonephritis ECG and BCG abnormalities of major severity were more common [6, 7].

2. The blood pressure in hereditary nephritis is generally normal. Seven of our patients were normotensive, 4 were moderately hypertensive. It was only in one case that the blood pressure attained 200/130 mm Hg.

According to our observations, the heart is not involved in this genetically determined process. As far as the heart is concerned, haemodialysis is thus most likely to be well tolerated in the terminal stage of the disease.

References

1. Alport, A. C.: Hereditary familial congenital haemorrhagic nephritis. *Brit. med. J. 1*, 504 (1927).
2. Antonovych, T. T., Deasy, P. F., Tina, L. U., D'Albora, J. B., Hollerman, C. E., Calcagno, P. L.: Hereditary nephritis: Early clinical, functional, and morphological studies. *Pediat. Res. 3*, 545 (1969).
3. Cassady, G., Brown, K., Cohen, M., DeMaria, W.: Hereditary renal dysfunction and deafness. *Pediatrics 35*, 967 (1965).
4. Crawford, M. D. A., Toghill, P. J.: Alport's syndrome of hereditary nephritis and diagnosis. *Quart. J. Med. 37*, 563 (1968).
5. Sessa, A., Cioffi, A., Conte, F., D'Amico, G.: Hereditary nephropathy with nerve deafness (Alport's syndrome). *Nephron 13*, 404 (1974).
6. Tarján, J., Balogh, Z., Gofman, L., Hámori, A.: Changes in the ballistocardiograms of nephropathic patients. Ballistocardiography and cardiovascular therapy. Proc. 2nd World Congr. Ballistocard. Cardiovasc. Dynamics, Oporto, 1969. *Bibl. cardiol.* Vol. 26, pp. 332–336. Karger, Basel–Munich–New York 1970.
7. Tarján, J., Balogh Z., Gofman, I., Hámori, A.: Ballistocardiographiás vizsgálatok nephropathiákban. *Magy. Belorv. Arch. 23*, 171 (1970).