RENAL DWARFISM

A RECORD OF ABNORMALITIES IN CARBOHYDRATE METABOLISM

BY

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Abnormalities have been observed in the carbohydrate metabolism of three of the four cases of renal dwarfism summarized below. Since the abnormalities do not appear to have been noted in previous cases of renal dwarfism it is felt that a brief record of the findings is of interest.

Case summaries

(Only relevant points are recorded)

Case 1. Male, aged 8 years when first seen. Birth weight, 7 lb. Difficult labour lasting three days. Growth had been stunted since the age of 3 years. Thirst and polyuria were noted by the mother at the age of 4 years. When first examined (January, 1942) he showed height, 42 inches; weight, 38 lb. He was healthy and intelligent in appearance. The limb-trunk ratio was normal. Blood pressure was 110/70 mm. Hg. The clinical examination was negative. The urine was pale, specific gravity 1002 to 1010, and contained no abnormal constituents. The twenty-four-hour volume was 60 to 110 oz. The x-ray of skull showed an unusually massive dorsum sellae and posterior clinoid processes. An excretion urogram (14 c.c. of uropac) showed no dye in pelves, ureters or bladder. Active rickets (type A) was first noted radiologically in November, 1942. The urine was tested daily from January to May, 1942, and for short periods on several subsequent occasions. No albumin found apart from a trace on two occasions. When last seen in November, 1943, however, the urine contained a cloud of albumin and the blood pressure was 155/120 mm. Hg (the first abnormal reading noted). Walking had deteriorated and the rickets had become more severe. He died in March, 1944: no details of death were available.

BLOOD CHEMISTRY (mgm. per cent.)

Date	Blood urea	Blood creatinin	Serum phosphorus	Serum calcium
April, 1942 ..	52	—	—	—
May, 1942 ..	109	—	4.4	9.5
June, 1942 ..	153	7.5	6.2	9.8
Aug., 1942 ..	129	—	4.8	9.8
Nov., 1942 ..	160	—	—	—
Dec., 1942 ..	270	3.2	—	—
Feb., 1943 ..	192	3.4	5.2	9.2
Nov., 1943 ..	189	—	5.7	8.5

Blood phosphatase activity normal.

Plasma proteins normal.

CO₂ combining capacity of plasma 30.9 c.c.

Urea concentration test (April, 1942) 10 gm. urea given.

Before urea ..	..	..	0.3 per cent.
1 hr. after urea ..	..	..	0.4 „ „
2 hr. after urea ..	..	..	0.4 „ „
3 hr. after urea ..	..	..	0.5 „ „

Daily blood sugars while in hospital were within normal limits.

Renal threshold for glucose=approx. 250 mgm. per cent. glucose.

INSULIN SENSITIVITY: 20.4 gm. glucose by mouth; 3.4 units soluble insulin i.v.

Fasting blood sugar ..	93 mgm. per cent.
10 min. after injection ..	94 „ „ „
20 min. after injection ..	87 „ „ „
30 min. after injection ..	86 „ „ „
45 min. after injection ..	93 „ „ „
60 min. after injection ..	100 „ „ „

TABLE 1—GLUCOSE TOLERANCE TESTS
(mgm. glucose per 100 c.c. of blood)

DATE ..	Feb. 3, 1942	Feb. 30, 1942	Mar. 19, 1942	June 17, 1942	July 23, 1942	Nov. 24, 1942	Nov. 10, 1943
DOSE ..	20 gm.	50 gm.	20 gm.	20 gm.	50 gm.	50 gm.	50 gm.
	Urine sugar	Urine sugar	Urine sugar	Urine sugar	Urine sugar	Urine sugar	Urine sugar
Fasting blood sugar ..	104 0	92 0	78 0	91 0	88 0	71 0	99 0
Glucose given							
½ hr. after ..	134 0	152 0	134 0	— 0	159 0	105 0	192 0
1 hr. after ..	202 0	243 0	174 0	169 0	207 0	158 0	309 Trace
1½ hr. after ..	258 0	239 Trace	179 0	115 0	293 Trace	221 0	378 —
2 hr. after ..	298 0	201 Trace	127 0	115 0	305 —	278 —	384 —
2½ hr. after ..	174 0	205 0	106 0	70 0	286 —	337 —	378 —

Case 2. Female, aged 3 years when first seen. Her mother noted the thirst when the patient was just over twelve months old. When first examined (Dec., 1941) she was a backward child. Her height was 31 inches, and weight 23 lb. The trunk-limb ratio normal. Her blood pressure was 105/70 mm. Hg. The clinical examination was negative. Her urine was pale, specific gravity 1002-1016, and contained no abnormal constituents. The x-ray of the skull was normal. No radiological evidence of rickets noted. A plain renal x-ray showed no opaque shadows; kidney shadows not outlined. On normal diet the blood sugar reached its maximum half an hour after mid-day (200 to 230 mgm. per cent. of glucose). No sugar was found in urine after blood specimens taken. Daily urine tests over a period of three months and during several short subsequent periods showed no albumin apart from a trace on one occasion. The case was last reviewed in November, 1943. Her height was 34½ inches and weight 29½ lb. Her blood pressure was 110/70 mm. Hg. The urine contained no albumin. No radiological evidence of rickets traced.

BLOOD CHEMISTRY
(mgm. per 100 c.c.)

Date	Blood urea	Blood creatinin	Serum calcium	Serum phosphorus
Jan., 1942 ..	27	—	—	—
Dec., 1942 ..	72	1.5	11.5	4.7
May, 1943 ..	44	—	—	—
Nov., 1943 ..	35	—	11.3	3.6

Blood phosphatase activity normal.

Urea concentration test (Dec., 1942) 4 gm. urea given.

Before urea	0.7 per cent.
1 hr. after urea	0.9
2 hr. after urea	0.5
3 hr. after urea	0.9

Urea clearance=52 per cent. average normal function (blood urea=71 mgm. per cent.).

Creatinine clearance, filtration per minute=78 c.c.

Insulin sensitivity=normal.

Renal threshold for glucose=approx. 250 mgm. glucose. See table 2 for glucose tolerance.

Case 3. Male, aged 7 years. Thirst was first noted when he was twelve months old. He was never very active. Generalized oedema gradually

developed in October, 1942. He was treated at another hospital for nephritis and bronchitis for three months during which period the oedema gradually subsided. Thirst and polyuria had been marked since. When first examined (July, 1943) he was a spare, intelligent boy, with fallow skin. His height was 41 inches, weight 34 lb. Trunk-limb ratio was normal. His blood pressure was 112/70 mm. Hg. Clinical examination was negative apart from evidence of bronchitis. His urine was pale, specific gravity 1008 to 1016, and twenty-four-hour volume 14 to 41 oz. Albumin (—) was found. The centrifuged deposit showed 8 red blood cells per field, with occasional pus cells, and a few granular casts. An excretion urogram (12 c.c. uropac) showed no dye in pelvis, ureters or bladder. The kidney shadows were small in size. Daily blood sugars half an hour after lunch ranged from 204 to 262 mgm. per cent.

BLOOD CHEMISTRY
(mgm. per cent.)

Date	Blood urea	Blood creatinin	Serum calcium	Serum phosphorus
July, 1943 ..	165	4.8	9.3	8.9

Urea clearance=6.5 per cent. average normal function. (Blood urea 219 mgm. per cent.)

Plasma proteins 6.2 gm. per cent.

GLUCOSE TOLERANCE TEST

Date: July, 1943

Dose	50 gm.	Urine Sugar
Fasting blood sugar ..	69	0
½ hr. after glucose ..	153	0
1 hr. after glucose ..	161	0
1½ hr. after glucose ..	193	0
2 hr. after glucose ..	220	0
2½ hr. after glucose ..	261	Trace

Renal threshold for glucose=approx. 250 mgm. glucose.

Case 4. Female, aged 17 months. She had always been a wet baby, passing urine every quarter hour. There was marked nocturnal enuresis. She drank two pints of cow's milk daily and four cups of tea. On examination her height was 27 inches, weight 14 lb. 8 oz. She was able to toddle. The limb-trunk ratio was normal. Clinical examination was negative. Her urine was pale, with a trace of albumin but no other abnormal constituents. There was no radiological evidence of rickets or other bone

TABLE 2—GLUCOSE TOLERANCE TESTS (Case 2)
(mgm. glucose per 100 c.c. of blood)

DATE	Jan., 1942	Dec. 11, 1942	Dec. 22, 1942	Jan., 1943	May, 1943	Nov., 1943
DOSE	14 gm.	40 gm.	60 gm.	50 gm.	50 gm.	50 gm.
	Urine sugar	Urine sugar	Urine sugar	Urine sugar	Urine sugar	Urine sugar
Fasting blood sugar ..	84	71	—	73	86	84
Glucose given ..	0	0	0	0	0	0
½ hr. after	121	139	243	172	—	209
1 hr. after	108	150	300	224	213	375
			(child vomited)			
1½ hr. after	91	164	224	251	157	175
2 hr. after	87	187	132	197	139	139
2½ hr. after	—	164	—	143	90	139

FIG. 1

abnormalities. While in hospital the child drank large quantities of fluid and passed much urine, but it was impossible to estimate even approximately the twenty-four-hour volume. This child has a twin brother who is healthy and normal in every respect (fig. 1).

BLOOD CHEMISTRY
(mgm. per cent.)

Date	Blood urea	Serum calcium	Serum phosphorus
February, 1944	104	13.5	5.3

Blood phosphatase activity slightly raised.
Plasma proteins 6.3 gm. per cent.

GLUCOSE TOLERANCE TESTS

DATE	..	..	Feb. 15, 1944	Mar. 2, 1944
DOSE	..	..	11 gm.	20 gm.
Fasting blood sugar	..	..	91	—
½ hr. after glucose	..	..	128	110
1 hr. after glucose	..	..	142	135
1½ hr. after glucose	..	..	117	107
2 hr. after glucose	..	..	75	—
2½ hr. after glucose	..	..	71	—

Discussion

The purpose of this article is not to discuss the theories of pathogenesis of renal dwarfism but simply

to record the changes in carbohydrate metabolism which have been observed in three of the four cases studied. These cases show a raised renal threshold for sugar (approximately 250 mgm. per cent. glucose) and a variable degree of diminished carbohydrate tolerance. From the data available it has not been possible to draw any conclusions regarding the causation of these changes. It is of interest, however, to recall that Lander et al. (1925) obtained similar glucose curves and a raised renal threshold for glucose in adolescents and adults suffering from nephrosclerosis with little or no renal insufficiency and that Castaigne and Chaumerliac (1939) found that a high renal threshold for glucose is very common in chronic sclerosing nephritis.

Hagedorn and Jensen method was used for all blood sugar estimations. (Capillary blood specimens were taken.)

Thanks are due to Dr. H. T. Ashby for allowing the publication of case 2 and to Dr. M. Flowerday for her assistance with the first two cases.

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