CONGENITAL VALVULAR OBSTRUCTION OF THE URETHRA.

BY

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Since Langenbeck¹, in 1802, first described a case of congenital valvular obstruction of the urethra, similar cases have been recorded from time to time as post-mortem findings, but little attention has been paid to the condition from the clinical aspect.

To Professor Young² of Johns Hopkins Hospital belongs the credit from making the first clinical diagnosis of valvular obstruction, and successfully operating on the patient, a boy of 17. The case was recorded at the Johns Hopkins Hospital Medical Society in 1913. In 1919 Young, Frontz, and Baldwin³ reported twelve cases in eight of which operation was carried out successfully, and recently Young and McKay⁴ have published a further account of the condition with a total of twenty-one cases, of which fifteen were successfully operated on at the Brady Urological Institute, Johns Hopkins Hospital.

My attention was first called to this condition by seeing the specimen from Dr. Hutchison's patient reported by Poynton and Sheldon⁵. This specimen led me to make a diagnosis in the cases reported below, one of them (Case 1) having been mentioned in the article by Poynton and Sheldon.

Professor Young³ describes three types of valve (Fig. 1).

Type 1. Ridge lying on the floor of the urethra continuous with the verumontanum, which takes an anterior course and divides into two fork-like processes in the region of the bulbo-membranous junction. These processes are continued as thin membranous sheets directed upward and forward, and may be attached to the urethra throughout its entire circumference. In the majority of cases of the general type the fusion of the valves anteriorly is not complete, there existing at the front a slight separation of the folds.

Type 2. In the second general type, there occurs a more or less cylindrical ridge similar to that found in the preceding type, with the exception that it passes from the upper aspect of the verumontanum towards the internal sphincter, where it divides into two fork-like processes which are continued as membranous sheets attached to the urethra just outside the internal sphincter in a manner similar to that described in the preceding type.

Type 3. There is a third type which has been found at different levels of the posterior urethra and which apparently bears no such relation to the verumontanum as the types just described. This was first mentioned by Jarjavay⁶ who described it as an iris valve. This was attached to the entire circumference of the urethra, there being a small opening in the centre. Incomplete varieties of this type have been described, the most common being a more or less crescentic or semicircular fold crossing the urethra and being attached either to the roof or floor.

Fig. 1. A diagram showing the three types of congenital valves of the posterior urethra. Type 1: a, Two bifurcated valves springing from distal portion of verumontanum. b, Two fused valves in same position. c, A unilateral valve in same position. Type 2: A bifurcated valve extending from proximal portion of verumontanum to lateral sides of prostatic urethra and roof. Type 3: a, Iris valve below verumontanum. b, Iris valve above verumontanum. The shaded circles represent the cystoscopic field seen in the region of valve. The internal sphincter and the prostatic urethra are shown to be dilated and the region of the membranous urethra is indicated. (By courtesy of Prof. Young from *Surg. Gyn. and Obstet.*, 1929, XLVIII, 517).

AUTHOR'S CASES.

Case 1. S. W., a boy originally admitted under Dr. Poynton in January, 1926, age 2 years and 10 months. The case has been described in the Archives of Disease in Childhood⁵. He was admitted for wasting. He had always been delicate and complained of great thirst.

On examination the bladder was found to reach up to the umbilicus, and there was constant enuresis; the urine was acid, contained a cloud of albumen and much pus, and on culture grew *Bacillus coli*. Blood urea=104 mgrm. per 100 c.cm. On cystoscopy the bladder was seen to be sacculated and trabeculated, and showed signs of chronic cystitis. There was no dilatation of the ureteric orifices. As there seemed to be some obstruction about the position of the compressor, bougies were passed and the urethra dilated as much as possible. Patient left the hospital very little improved.

He was re-admitted to hospital in June, 1927, in the same condition as in 1926. The bladder was up to the umbilicus and the urine continuously dribbling away.

Blood urea	...	...	60 mgrm. per 100 c.cm.
Urea concentration, 24 hours	...	...	85%
After urea 1st hour	...	...	1.08
2nd hour	...	...	2.45
3rd hour	...	...	0.05

The urine contained pus and albumen and gave a profuse growth of *B. coli*.

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On June 26th the bladder was opened and with a head light a good view of the internal meatus was obtained. The internal meatus and prostatic urethra were seen to be dilated, but it was not possible to see the whole length of the prostatic urethra. No obstruction was seen. Bougies were passed through the internal meatus, and the urethra dilated as much as possible. The wound healed quickly except that an intermittent supra-pubic fistula formed, in spite of which patient was able to pass water naturally, with a good stream.

In January, 1928, patient was re-admitted for closure of the fistula. The fistula was excised and the opportunity taken once more to dilate the urethra by passing bougies *via* the internal meatus. The supra-pubic wound was slow to close but ultimately healed soundly, and patient was able to pass a good stream of urine.

After leaving hospital the boy's general health improved considerably and his thirst became much less, but he still remained very pale and did not appear to be very strong.

In November, 1928, he was re-admitted for tonsillectomy and the opportunity was taken to examine the condition of the bladder and kidneys. Urine was passed with a good stream but there were four ounces residual urine. A profuse growth of *B. coli* was obtained. Blood urea examination done on three occasions varied from 74 to 110 mgrm. per 100 c.cm.

Urea concentration test:

			Urea %		Vol. c.cm.
Before urea	...	...	1.52	...	—
After urea 1st hour	...	...	1.50	...	44
2nd hour	...	...	1.45	...	65
3rd hour	...	...	1.35	...	70

On cystoscopy the bladder showed signs of mild chronic cystitis. There was much trabeculation and sacculation. The ureteric openings were not obviously dilated but no contractions were seen. Catheters were passed up both ureters and 10 c.cm. of sodium iodide was injected into each. The pyelogram showed the catheters curled up in enormously distended ureters and a high degree of distension of the pelvis and calyces of both kidneys (Fig. 2).

In June, 1929, the boy's height and weight were 3ft. 7½ in., and 34lb. respectively at the age of 6 years, 4 months.

Case 2. A. G., male, aged 8 years and 8 months, admitted to hospital, October, 1927.

History of bladder trouble for four years getting worse for four or five months. Straining on trying to pass water, poor stream, water dribbling away. Frequency during day five minutes to three hours. Nocturnal incontinence.

The boy's general condition seemed to be pretty good though he was rather pale. The bladder was distended and reached up to the umbilicus. The kidneys could not be felt and there was no tenderness in the loins. On attempting to micturate the urine only trickled away. The urine on examination showed a trace of albumen, many pus cells and a profuse growth of *B. coli*. Sp. Gr.=1020. Blood urea=30 mgrm. per 100 c.cm.

Urea concentration test:

			Urea %		Urine c.cm.
1st hour	...	...	1.05	...	148
2nd hour	...	...	0.70	...	137
3rd hour	...	...	1.15	...	50

On October 20th through a mid-line supra-pubic incision the bladder was exposed and stripped down from the back of the pubis. The prostatic urethra was exposed beneath the pubic arch and opened. A good view of the prostatic urethra was obtained but no valves or obstructions were seen. The veru montanum appeared to be very large and inflamed, and was touched with the cautery, as it was thought possible that it might have caused a reflex spasm of the compressor. Bougies were passed through the opening and down the urethra. The opening in the urethra was closed and a drain placed in the bladder. With daily irrigations of the bladder the boy's general condition improved greatly and in March the urea concentration test showed:—

			Urea %		Urine c.cm.
24 hours urine	...	...	2.41		
1st hour	...	...	2.31	...	71
2nd hour	...	...	2.72	...	68
3rd hour	...	...	2.90	...	85

Unfortunately a supra-pubic fistula formed and showed no signs of closing. On May 10th, 1928, the fistulous track was separated with great difficulty from the back of the pubis and followed to the prostatic urethra. The track was excised and the opening in the urethra closed. The fundus of the bladder was opened and the urethra was once more dilated as fully as possible by bougies passed through the internal meatus. A drainage tube was put into the bladder, and the space in front of the bladder down to the urethra was packed with gauze soaked in flavine to allow the opening in the urethra to close securely before becoming adherent to the pubis. The wound healed without trouble and the boy left the hospital able to pass a good stream of urine.

He was re-admitted to hospital in April, 1929, for investigation of renal functions, etc. The general condition of the patient was greatly improved, the supra-pubic scar was sound, and he was able to pass a normal stream of urine.

The urine was acid sp.gr. 1020–1028 and still heavily infected with *B. coli*. Blood urea was 39 mgm. per 100 c.cm.

Urea concentration test :

				Urea %		Volume c.cm.
Before urea	...	...	...	3.71		
1st hour	...	...	...	2.91	...	42
2nd hour	...	...	...	2.76	...	58
3rd hour	...	...	...	2.64	...	29

Cystoscopic examination showed much trabeculation and sacculation, very slight evidence of chronic cystitis. Both ureteric orifices gaping a little. Pyelography with sodium iodide showed slight degree of hydronephrosis of both sides (Fig. 3).

Case 3. N. J., male; aged 9½ years, admitted to hospital October 20th, 1928, with a history of incontinence of urine day and night since birth. The water has always been quite thick and foul smelling; thirst very troublesome for years. Has had no headache, nor vomiting, and has not complained of pain, but appetite is very poor and he seems to be losing weight.

The boy's general condition appeared to be fairly good. The bladder was up to the umbilicus and there was some tenderness over the right kidney, but neither kidney could be felt. On attempting to pass urine only a fine trickle escaped from the penis, and twenty ounces residual urine were found on passing a catheter. The urine was acid and contained stringy muco-pus, and a trace of albumen; it was infected with *B. coli*. Blood urea=89 mgm. per 100 c.cm. The urea concentration test showed :

				Urea %		Vol. c.cm.
Before urea	...	...	...	0.85	...	—
1st hour	...	...	...	0.91	...	43
2nd hour	...	...	...	0.90	...	59
3rd hour	...	...	...	0.89	...	78

X-ray of the renal tract was negative.

On October 25th cystoscopy was performed. A slight but definite obstruction to the passage of the cystoscope was felt about the position of the compressor. It was found impossible to wash the bladder quite clean, owing to the amount of pus collected in the dilated ureters and kidneys. The bladder was greatly trabeculated and sacculated, and the opening of the ureters were much dilated; no contractions of the ureters were seen. A small bougie was tied in the urethra with the idea of gradually emptying the bladder. Patient had a rigor the same night.

As the urethra quickly became intolerant of a bougie, a De Pezzer catheter was put into the bladder above the pubes on November 1st. The bladder was then washed out daily, the urine becoming considerably cleaner. When the bladder was washed out through the supra-pubic catheter a good stream of fluid passed through the urethra. On November 15th the urethra was examined through a special irrigating urethroscope, size No. 9 Charrière made by the Genito-Urinary company. It was found impossible to obtain a clear medium but a view was obtained of a greatly dilated and very inflamed posterior urethra. No fold or valve was seen but two openings of some size were seen in the floor of the urethra some distance on the distal side of the veru montanum. In spite of continued daily irrigation of the bladder the patient became uraemic and died on November 27th, 1928.

Only a partial post-mortem examination through the supra-pubic wound was permitted. All that was done was to remove the whole genito-urinary tract intact from kidneys to urethra. Both kidneys were soft, flabby, and bag-like. Ureters both dilated and thickened. Bladder greatly hypertrophied, with supra-pubic opening. Specimen hardened in formalin before further examination and therefore the figure shows much less than the actual dilatation (Fig. 4 and 5).

Further examination :—Bilateral hydronephrosis; grossly dilated ureters. Ureteric orifices into bladder on the small side. Bladder very hypertrophied.

Urethra :—Veru montanum very big (size of small pea) and just in front of it appears to be a valve like obstruction—almost a double canal to the urethra about $\frac{1}{3}$ -in. in length. In front of this again the urethra is dilated and shows signs of inflammation of its mucosa. Two openings which would admit a small probe were seen in the floor of the urethra and were thought to be the openings of the ducts from Cowper's glands.

The valve-like fold in the prostatic urethra can hardly have been the cause of the obstruction since the urethra was dilated for a considerable distance on the distal side of it, and it seems likely that the actual cause was a partial diaphragm distal to the compressor. This diaphragm was destroyed by the passage of the cystoscope. An obstruction in this situation would account for the paralysis of the compressor which allowed fluid to escape by the urethra while the bladder was being washed out through the supra-pubic opening.

DISCUSSION.

Retention of urine in a child is usually due to phimosis, a pinhole meatus, some local inflammation, or sometimes to a calculus impacted in the urethra.

In the absence of one of these causes, the diagnosis of valvular obstruction can safely be made in any case where the signs are those of chronic retention and overflow, with a history dating back to infancy. There is always difficulty of micturition from birth, and the stream is very weak, or a mere dribble. Later continuous enuresis develops. In the more severe grades of obstruction the back pressure effects on the kidneys rapidly lead to a fatal result, as in the case recorded by King and Sheldon⁷, but in cases showing the lesser degrees adult life may be reached. Usually the patient is brought for treatment during infancy or early childhood. On examination the child is found to be small and undeveloped for his age, generally pale and listless, often very thirsty, and showing signs of chronic uræmia. The bladder is distended and reaches up to the umbilicus or higher, urine is constantly dribbling from the penis, and the dilated ureters may sometimes be felt through the abdominal wall. The urine is of low specific gravity, contains a cloud of albumen and is always badly infected, usually with bacillus coli. Renal function tests will show diminished kidney activity. On the passage of an instrument through the urethra, the obstruction if due to a diaphragm (3rd type) may be recognized; but if due to a valve (1st or 2nd type), which will lie flat against the wall of the urethra, nothing abnormal will be felt. On cystoscopy the bladder shows signs of chronic cystitis and the usual evidence of back pressure in the form of trabeculation and sacculation. The openings of the ureters will be gaping to a greater or less degree and usually motionless.

The simplest method of treatment seems to be the retrograde passage of bougies, that is from the bladder towards the meatus, and in most cases this will probably effect a cure. Professor Young has invented a special punch by means of which he removes the valve after locating it with an urethroscope,

but the great degree of skill necessary to use such an instrument in the urethra of an infant must require an amount of practice that the ordinary surgeon is unlikely to obtain.

In the first case on which I operated (Case I) and passed bougies from the bladder, I think it doubtful whether I completely destroyed the valve, for the boy developed a supra-pubic fistula which did not heal until the bladder had been re-opened and bougies again passed. On account of this partial failure, in treating Case 2 I cut down on the urethra, and although I obtained a good view of the prostatic urethra I could see no valve. However the passage of bougies has cured the boy and he now passes an absolutely normal stream.

I have seen four patients with this condition within the last three years, and also two post mortem specimens, so that the condition cannot be so rare as is generally supposed. I remember on more than one occasion in the past having seen similar cases which were undiagnosed, and now that attention is being called to the condition I have no doubt that many more will be discovered before long.

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Fig. 2. (Case 1). Double pyelogram Nov., 1928. Pelves and calyces of both kidneys greatly dilated. Catheters curled up in largely distended ureters.

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Fig. 3. (Case 2). Double pyclogram, April, 1929, showing slight degree of hydronephrosis and dilatation of ureters on both sides.

Fig. 4. (Case 3). Kidneys, bladder and urethra showing destruction of kidneys, dilatation of ureters, and great hypertrophy of bladder.

Fig. 5. (Case 3). An enlarged view of part of the urethra and bladder in Fig. 4. The urethra was dilated as far as the distal end of the rod passing beneath the fold in the floor.