STENOSIS OF THE URETEROPELVIC JUNCTURE: CONGENITAL AND ACQUIRED

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Conditions which may produce obstruction at the upper end of the ureter and cause hydronephrosis are manifold. In many instances a combination of these is present, creating a situation of such complexity that the primary cause of the obstruction may not be recognized at operation. This may lead to an improper or inadequate surgical procedure in which the underlying cause is not removed. The hydronephrosis therefore naturally persists.

During the past 2 years a study has been made of these upper ureteral obstructions to determine which, if any, were characteristically primary, and which were superimposed conditions acquired during the progress of the dilatation. The material upon which this investigation was based consisted of 71 cases of hydronephrosis in the Brady Urological Institute that were operated upon, excluding cases of obstruction caused by calculus, tumor, or tuberculosis. I am indebted to Dr. Hugh H. Young for permission to study this material.

Of these 71 cases of hydronephrosis resulting from obstruction at the upper end of the ureter, there were only 4 cases in which the operator felt that bands and kinks were the primary cause. In 24 instances vessels were found to be responsible. A survey of this group, together with an interpretation of the mechanisms by which these vessels may exert their obstructive contact, has been made in a previous publication. It was the purpose of this study to determine the nature and significance of stenosis of the ureteropelvic juncture, which caused the hydronephrosis in the remaining 43 cases.

In 10 of these 43 cases the stenosis was associated with accessory vessels at the lower pole of the kidney (fig. 1). In this group, however, the stenosis was considered primary for 5 reasons. First, stenosis without accessory vessels was encountered more than 3 times as often as stenosis with accessory vessels. Second, the average age of onset of symptoms in this group was 13 years, as compared with 24 years in the group of accessory renal vessels without associated stenosis. Third, the duration of symptoms in this group was only one-fourth as long as it

was in the group with vessels alone, a fact which made it seem unlikely that contact was productive of stricture. Fourth, vascular obstruction was unassociated with stenosis in 24 instances. Finally, the stenosis itself was delicate and thin and showed no evidence grossly of changes which might have been induced by trauma or inflammation.

There were 33 cases of stenosis of the upper end of the ureter without vessels. Sixteen of these showed inflammatory changes, and had

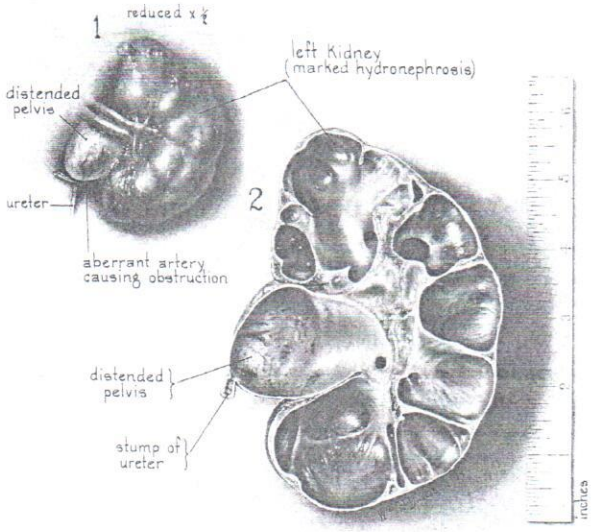

FIG. 1. Drawing of a case of congenital stenosis of the ureteropelvic juncture associated with accessory renal vessel.

infected urine. In 7 an inflammatory reaction was proved by sections. This reaction was characterized in some instances by cellular infiltration and oedema in the submucosa, in others by subepithelial fibrosis, and in still others by scarring in the muscle or adventitia. In 9 of these 16 cases, histological sections were not available; but on gross examination there was evidence of a fibrous thickening at the site of the stenosis.

In 17 of the 33 cases of stenosis without vessels, there was no evidence of an underlying inflammatory reaction (fig. 2). Sections were

available for microscopic study in 4 of these cases (fig. 3). In these the stenosis was found to be thin and delicate. The circumference of the ureter at the site of the stenosis was considerably reduced; and the diameter of the lumen, as well as the external diameter, was also much

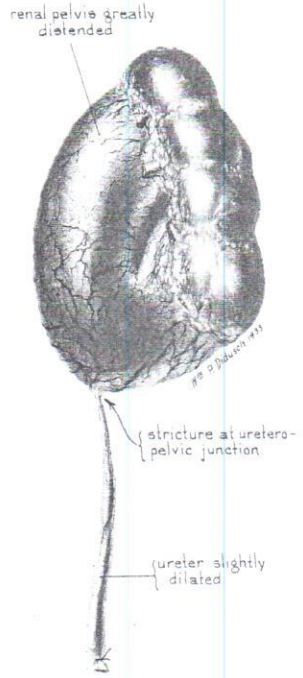

FIG. 2. Drawing of a case of congenital stenosis of the ureteropelvic juncture without associated vessels.

shorter than the corresponding dimensions in the normal ureter below. In 13 cases sections were not available for study, but detailed gross descriptions justify us in including them in the same group. Eleven of these 17 cases had uninfected urine.

Inasmuch as no acquired factors could be found to explain this delicate type of stenosis, it seemed evident that it must have been of congenital origin. Two explanations for such an etiology presented themselves. Either some factor in embryonic life actively interfered with the development of the upper end of the ureter; or else a sufficient stimulus for its growth was lacking. Felix stated that, after the tip of the ureter had attained its definitive position opposite the second lumbar vertebra in embryos of 9.5–13 mm. greatest length, the ureter must elongate enormously, since it is in the region of the lumbar vertebrae that the body growth principally takes place. Conceivably, if the body growth,

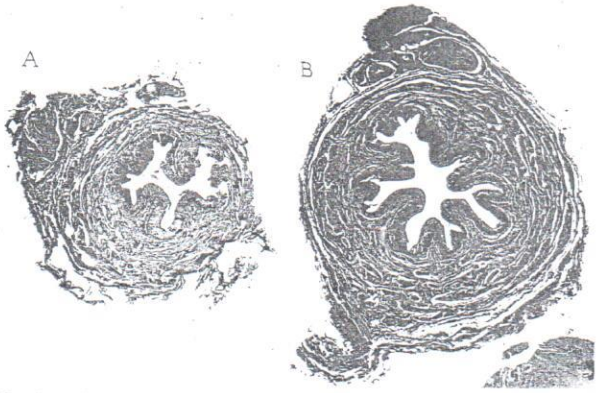

FIG. 3. *A*, Transverse section through congenital stenosis which caused hydronephrosis. *B*, Section through normal ureter in same case 5 mm. below. No inflammation is present. The muscle coat of the normal ureter is slightly thicker than that of the stenotic ureteropelvic juncture. B. U. I. Path. No. 9481.

in any given case, is more pronounced in the lumbar region below the level of the ureteropelvic juncture, there may be an insufficient stimulus for the proper development of the upper end of the ureter.

A preliminary study of serial sections of 11 embryos in the Embryological Department of the Carnegie Institution of Washington, under the guidance of Dr. George L. Streeter, has failed so far to disclose any peculiarities at the ureteropelvic juncture which might be a factor in the production of a congenital stenosis. The muscle coat of the pelvis and that of the upper ureter apparently are laid down simultaneously in embryos which have attained a crown-rump length of 90 mm. In these,

as well as in larger embryos, the ureter expands so gradually into the pelvis that it is impossible to determine where one ends and the other begins. The epithelium, as well as the muscle coat, appears to be the

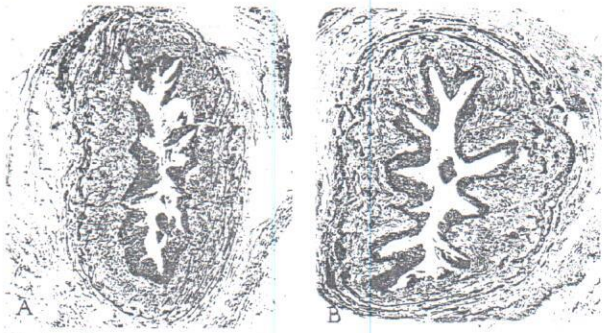

FIG. 4. *A*, Transverse section through a well-defined uretero-pelvic juncture in a stillborn infant. There is no suggestion of a pyeloureteral sphincter. *B*, Section through the normal ureter in same case 1 cm. below. The muscle coat here is much better developed than in *A*. No hydronephrosis. Gen. Path. No. 16599.

FIG. 5 FIG. 6 FIG. 7

FIG. 5. Persistent ureteropelvic juncture; 14 per cent of the cases studied. B. U. I. x-ray No. 22536.

FIG. 6. Normal, funnel-shaped, pyeloureteral outlet; 86 per cent of the cases studied. B. U. I. x-ray No. 22555.

FIG. 7. Transitory contraction phase at pyeloureteral outlet suggesting ureteropelvic juncture, in same case as figure 6. B. U. I. x-ray No. 22555.

same in pelvis and ureter; and nowhere can there be found any increase in the circular muscle fibers which might be interpreted as representing a pyeloureteral sphincter.

In view of the fact that Felix stated that the upper end of the ureter was narrow in all embryos whose length exceeded 125 mm., a study was

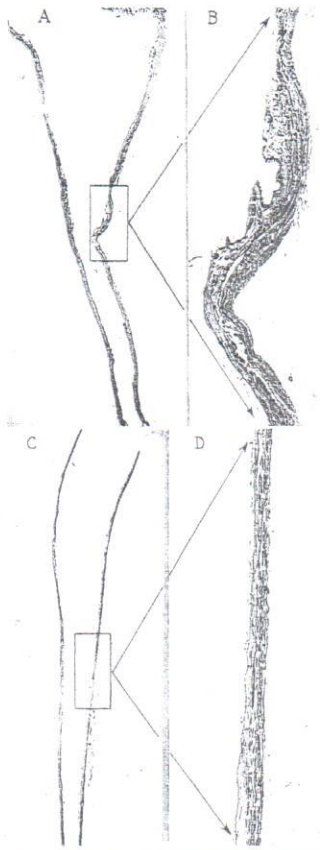

FIG. 8. *A*, Longitudinal section through well-defined ureteropelvic juncture removed from an adult at autopsy. No hydronephrosis. Gen. Path. No. 16784. *B*, Same. Higher magnification of region of ureteropelvic juncture. No pyeloureteral sphincter. *C*, Longitudinal section through a normal, funnel-shaped, pyeloureteral outlet. (Gen. Path. No. 16754.) *D*, Same. Higher magnification of region of pyeloureteral outlet, showing absence of pyeloureteral sphincter.

made of 50 upper urinary tracts removed from stillborn cases at autopsy. Only 8 (16 per cent) of these were found to have a ureteropelvic juncture (fig. 4). Forty-two (84 per cent) had a funnel-shaped pyeloureteral outlet. These findings were corroborated by a study of 200 consecutive normal urograms (figs. 5, 6, 7). Of these only 27 (14 per cent) showed a persistent ureteropelvic juncture. One hundred and seventy-three (86 per cent) showed a gradual, funnel-shaped pyeloureteral outlet with no discernible line of demarcation between pelvis and ureter (fig. 8). Without placing too much emphasis upon what may be a mere coincidence, it seems evident that in the majority of cases a true ureteropelvic juncture does not exist. It is possible, therefore, that a ureteropelvic juncture represents a minimal narrowing which, when present in marked degree, becomes a congenital stenosis.

In the group of cases comprising inflammatory strictures, it is impossible to determine with any degree of certainty whether the inflammatory reaction was primary or whether it was superimposed upon a simple congenital stenosis. It is obvious that necrosis of the ureteral wall from infection may be followed by so much scarring that contracture may occur. But many cases of chronic pyelonephritis are observed at autopsy with no evidence whatever of contracture at the ureteropelvic juncture, although microscopic sections show an advanced degree of scarring in the wall of pelvis and ureter. The fact that most nonspecific and non-traumatic inflammatory strictures affect the same site as that of a congenital stenosis suggests, at least, that an unrecognized congenital stenosis may be present first. Furthermore, in many instances, the same inflammatory change may be seen in sections taken from the ureter well below the stricture, with narrowing only at the ureteropelvic juncture. It seems likely then that infection is more apt to produce a fibrous stricture when superimposed upon a congenital stenosis or upon a well-defined ureteropelvic juncture.

Development of hydronephrosis from stenosis. The pyelectasis is influenced by the caliber of the pyeloureteral lumen, accessory vessels, infection, kink and fixation, high ureteral insertion, and, possibly, by renal growth.

Caliber: In the 11 cases of simple congenital stenoses, uncomplicated by infection or accessory vessels, the ages varied from 6 to 53 years. One stillborn at autopsy was found to have a congenital stenosis with hydronephrosis. Since the dilatation of the several renal pelves in this group occurred with such varying rapidity, it may be reasonably as-

sumed that such a result is attributable chiefly to a variation in the caliber of the different pyeloureteral lumens.

Vessels: In 8 of the 10 cases of congenital stenosis associated with accessory lower polar vessels, in which there was no infection, there were 4 patients who were 6 years of age or younger. The average age of all patients in this group was 14.6 years, as compared with 24 years in the group of uncomplicated congenital stenoses. This would seem to indicate that the development of hydronephrosis is accelerated by the presence of an accessory vessel when contact is finally established by the expanding pelvis.

Infection: The fact that the patients in this group were somewhat older than those in the group of uninfected congenital stenoses suggests merely that sterile cases sooner or later have a tendency to become infected. It is obvious that if an infection were superimposed upon a simple congenital stenosis, as it must be in many instances, it would reduce still further the caliber of the lumen and thus serve to accelerate the pyelectasis.

Kink and Fixation: This condition in the absence of vessels or bands may arise from an asymmetrical dilatation of the pelvis, elongation of the ureter from a low obstruction, ptosis, or peri-ureteral inflammation. It obviously increases the obstruction and hastens the dilatation.

High Insertion: This results from advanced asymmetrical dilatation of the renal pelvis, and may follow any of the above-mentioned obstructive factors. A case of high insertion unassociated with an underlying obstruction has never been observed in the Brady Urological Institute.

Renal Growth: Felix has said that the growth in length and thickness of the kidney takes place in periods. In intra-uterine life, it attains a length of 50 mm. During the first year it increases in length to 70 mm. From the second year onward it finally reaches a length of 120 mm., and most of this growth occurs during puberty. A somewhat narrowed ureteropelvic juncture, though adequate for an infantile kidney, may prove insufficiently patent when full maturity is reached.

Stenosis of wide caliber. When stenosis at the ureteropelvic juncture is of marked degree, it is easily detected at operation. The problem is merely selection of the most suitable procedure for the correction of the obstruction which has been identified. We have seen, however, that the caliber of the lumen may vary, and it is the stenoses of wide caliber that may remain unrecognized. In some of these instances the operator erroneously attributes the obstruction to a conspicuous

but secondary, superimposed factor, such as a kink or band; or, when this is absent, to some obscure dysfunction of the neuromuscular mechanism, because gross examination of the upper end of the ureter fails to reveal any definite stenosis. In order to appreciate the significance of a stenosis of wide caliber as a cause of obstruction, one must understand the sequence of stages through which pelvic dilatation progresses. This is illustrated in figure 9. At first the musculature of the renal pelvis hypertrophies (fig. 9, *3*) in order to contract more forcibly. So long as it

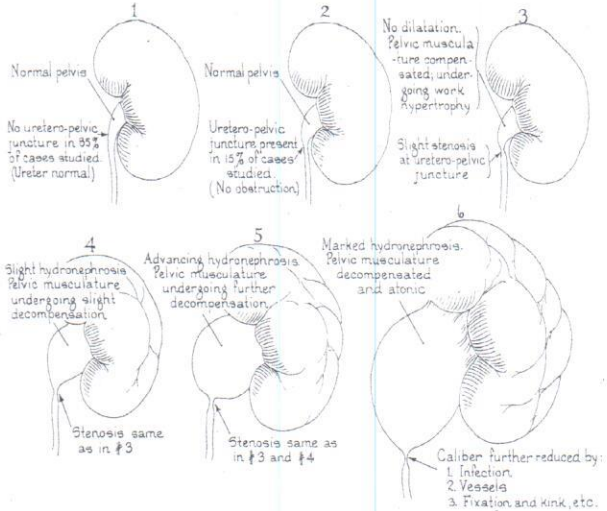

FIG. 9. Diagrammatic representation of the effect upon the renal pelvis of a stenosis of moderately wide caliber.

is able to function efficiently no dilatation occurs, although the strong, often spasmodic, contractions sometimes cause pain. The physiological principle of work hypertrophy of a muscular structure as a means of compensating for the increased effort placed upon it is well known. When its limit of endurance is reached and it can undergo no further hypertrophy, the pelvis begins to dilate. The expansion usually is gradual unless suddenly accelerated by the advent of such secondary factors as have been mentioned above. The less complete the obstruc-

tion, the better able the renal pelvis is to compensate. In its end stage the renal pelvis usually is a dilated, thin-walled, atonic sac (fig. 10).

In the group of simple congenital stenoses, uncomplicated by infection, vessels, or kinks and adhesions, the lumen in each case presumably had remained relatively constant in size after full maturity had been attained. Although the degree of stenosis must have varied somewhat among the several cases in this group, the variation, in actual measurements, could have been only slight; for its extremes were necessarily limited by reason of the small size of even a normal ureter. The

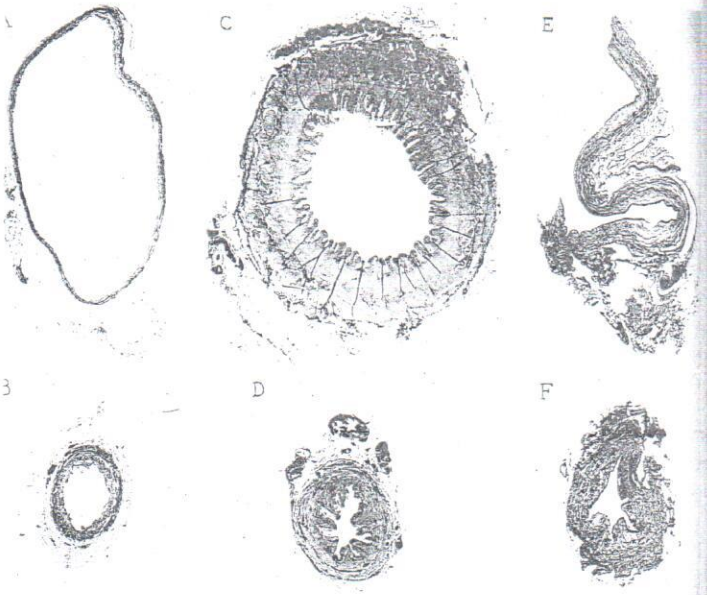

FIG. 10. *A*, Transverse section through normal renal pelvis. *B*, Same, through corresponding ureter. The muscle coat of the ureter is a little thicker. Gen. Path. No. 16754. *C*, Transverse section through renal pelvis 1 cm. above a stenotic ureteropelvic juncture. Marked hypertrophy of pelvic musculature. Hydronephrosis present and mostly intrarenal. *D*, The ureter 1 cm. below the stenosis. The muscle here is also thicker than normal. Path. No. 10861. *E*, Longitudinal section through dilated wall of renal pelvis in case of huge hydronephrosis. *F*, Transverse section through corresponding ureter 2 cm. below stenotic ureteropelvic juncture. B. U. I. Path. No. 6469.

variation in caliber, therefore, was not commensurate with the wide disparity in ages (from birth to 53 years). The ability of the musculature of the renal pelvis to undergo work hypertrophy and thus to function efficiently in the presence of obstruction is a factor almost equal in importance to the degree of stenosis. If the caliber of the lumen is gradually reduced, there must be a critical point at which the pelvic musculature is just able to compensate, and beyond which decompensation finally occurs and hydronephrosis develops. This critical point must vary in different individuals in accordance with the muscular development with which the renal pelvis is endowed and the ability of this musculature to undergo work hypertrophy. It is therefore unreasonable to dismiss as normal a pyeloureteral outlet of a very slightly reduced caliber merely because in another individual the same caliber is adequate. Inasmuch as some doubt now has been cast upon the existence of a point of physiological narrowing at the ureteropelvic juncture, any detectible narrowing, regardless of degree, should be viewed with suspicion when hydronephrosis is present. Achalasia, or failure of relaxation of the pyeloureteral outlet, though sometimes due to increased muscular tonicity, is probably more often the result of stenosis, which may be so slight that superficial examination fails to disclose it. The desirability of a plastic operation in the case of a minimal or questionable narrowing which is permanently persistent must be carefully considered, because in the majority of normal cases a line of demarcation between the pelvis and the ureter does not exist, and the pyeloureteral outlet is funnel-shaped.

I wish to express my appreciation to Dr. George L. Streeter, Dr. Arnold R. Rich, Dr. Richard W. Satterthwaite, and Dr. John S. Haines for their assistance in the preparation of this paper.

SUMMARY

A study of 71 cases of hydronephrosis has established 3 fundamental causes of obstruction at the upper end of the ureter: 1) bands and kinks, 4 cases (5.6 per cent); 2) accessory renal vessels, 24 cases (33.8 per cent); 3) stenosis, 43 cases (60.5 per cent). In the group of cases in which stenosis was the underlying cause of obstruction, secondary accelerating factors were accessory renal vessels, infection, kink and fixation, high ureteral insertion, and, possibly, rapid renal growth during puberty.

In the majority of normal cases there is no line of demarcation be-

tween the renal pelvis and the ureter. Any deviation from the normal funnel-shaped, pyeloureteral outlet is probably pathological. Deviation of a moderate degree, sufficient to cause only minimal obstruction, can be compensated for by work hypertrophy of the pelvic musculature.

When the ureter is normal, a sharply defined and permanently persistent ureteropelvic juncture, in the presence of pyelectasis, should be considered obstructive.

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MALIGNANT PAPILLARY CYSTADENOMA OF THE KIDNEY: CASE REPORT

WALTER GIFFORD HAYWARD

The case to be reported is of more interest because of its rarity than because of any practical benefit that can be derived from it. Search of the literature on the subject was rewarded with the finding of only a few case reports.

Case report. J. A., male, age 71, W. C. A. Hospital, Case No. 50755, gave the following history: With the exception of discomfort occasioned by the presence of varicose veins in both legs and a varicose ulcer on the right leg that appeared in the spring of 1939, the past history was entirely negative. He had always considered himself well and strong and had worked hard. There had been no urinary symptoms of any kind. Four days before admission, an aching pain had appeared in the chest that had been severe enough to cause cessation of work. He did not call a doctor but remained at home. After 2 days of the pain, he became nauseated and from that time until he entered the hospital, he was unable to retain food.

Physical examination: Pulse and temperature were normal. Blood pressure was 95/50. The abdomen was very large, flabby, and not tender. On palpation a rounded mass, approximately the size of a football, was discovered occupying the left side of the abdomen. Inferiorly, it extended below the anterior superior spine of the ilium; medially, it reached the midline; and superiorly, it disappeared under the costal margin and filled the flank. It was smooth, firm, immovable, and not tender. The patient had never been aware of its presence. The genitalia were normal. The prostate was normal in size and consistency. Varicose veins were present in both legs. The right leg was larger than the left. An indolent, thick-edged granulating ulcer was present on the anterior aspect of the lower third.

Laboratory Tests: Hemaglobin 61 per cent, red blood cells 3,910,000, white blood cells 16,800, polymorphonuclears 80 per cent, lymphocytes 17 per cent, monocytes 3 per cent.

Urinalysis: specific gravity 1.018, pH 5, albumin +, sugar negative, on microscopic examination a few finely granular casts, an occasional epithelial cell and a rare pus cell; non-protein nitrogen 32.4 mgm., urea nitrogen 11 mgm., sugar 133 mgm., creatinin 1.4 mgm., chlorides 360, occult blood, benzidine +, guaiac, trace.

Because of the presence of the huge abdominal tumor, a cystoscopic examination was requested. This was my first opportunity of seeing the patient.