Section of Urology

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Meeting October 27 1966

President's Address

Horseshoe Kidneys [*Abridged*]

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During the last forty years 9 cases of horseshoe kidney have been reported to this Section, 4 by Jocelyn Swan (1926), one by Dunhill (1932) and 4 by Robinson (1936).

The horseshoe kidney was originally regarded as a rare anatomical curiosity, which was seen only at post-mortem. When renal surgery was introduced some eighty years ago it was occasionally observed in the course of operation, although the surgeon usually closed the abdomen without touching the horseshoe. In this present age of diagnosis with the aid of retrograde pyelograms, intravenous urography and renal arteriograms, the incidence of horseshoe kidney is estimated at 1 in 200–400 individuals.

Embryology: The horseshoe anomaly results from an embryological fault which develops between the 4th and 8th week of intrauterine life. The renal blastemata become fused before rotation and migration: this fusion prevents independent rotation and the vessels thus develop an abnormal relation to the renal pelvis and ureters (Fig 1). The normal kidney in the course of development travels upward from near the level of the second sacral vertebra to the lumbar region. Horseshoe kidneys on the contrary are usually found to be ectopic, low in position and sited at the aortic bifurcation.

Anatomy: The shape of the symmetrical organ is that of a horseshoe, with a bridge of renal tissue uniting the lower poles across the middle line of the vertebral column (Fig 2). The posterior surface of the junction may be grooved by the aorta, which it may also compress. The junction may occur at the upper poles but this is rare and the isthmus will not necessarily be at the upper extremities: the organ may in fact take the form of an irregular 'H' (Fig 3), due perhaps to further ascent being prevented by the origin of the inferior mesenteric artery. In 1955 Culp & Winterringer reviewed the Mayo Clinic cases from 1912 to 1953: they found 106 cases with one fusion at the upper pole.

The connexion in a horseshoe kidney may be solid renal parenchyma or merely a fibrous band. The bridge may lie behind the inferior cava but this is rare. The ureter arises from the anterior surface of the pelvis: it tends to be obstructed if the pelvi-ureteric junction is high or if the ureter crosses over the prominent lower pole or

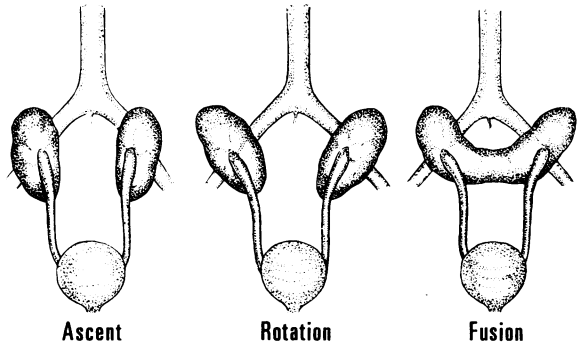

Fig 1 *The development of a horseshoe kidney*

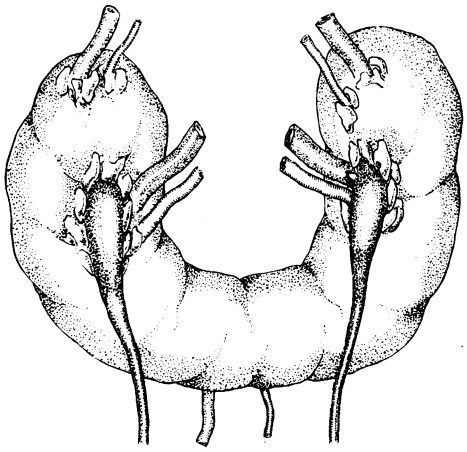

Fig 2 *Typical horseshoe kidney*

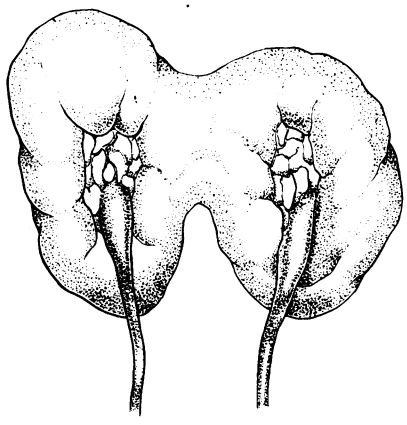

Fig 3 *Fusion near the upper pole, the irregular 'H'*

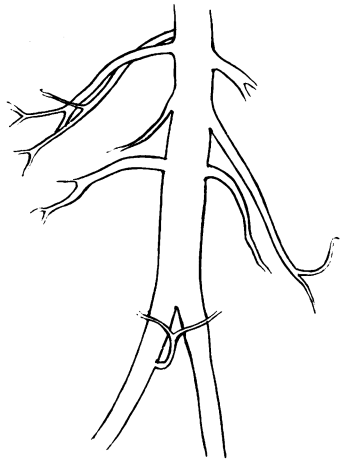

Fig 4 *Key diagram of aortogram, showing three arteries to each kidney and, arising from the right common iliac artery, an artery to the isthmus*

bulky isthmus. Anomalies in the blood vessels are common. There are frequently one, two or three arteries to each kidney arising from the aorta and a further one running to the upper or lower part of the isthmus. This latter artery arises from the aorta or from either of the common iliac arteries and divides to supply each half of the isthmus (Fig 4).

Pathology: Thompson (1929) collected 19 cases from post-mortem records, including two with upper polar fusion. He speaks of 'good lives', 'poor lives' and 'bad kidneys', and advances a plea for the organs to be examined *in situ* by a competent observer. Museum specimens are almost invariably found to have had all the arteries and veins removed.

About 25% of horseshoe kidneys are normal and give rise to no trouble (Glenn 1959): indeed, they are only revealed by accident, as for instance in an intravenous urogram performed before a prostatectomy. The danger nevertheless remains that they can lead to manifold complications. A high origin of the ureter from the pelvis or a ureter which crosses a lower pole or a prominent isthmus may well cause a hydronephrosis. The ureters do not show in an intravenous urogram, so that the pelves may fill up, with the result that urine spills down slowly into ureters which are seldom visible. Hydronephrosis develops with subsequent infection, pyelonephritis or pyonephrosis. The stagnation may lead to stone formation, often bilateral: moreover, after removal the stones recur with amazing rapidity. All these factors involve the patient in a remote risk of renal failure.

Injury: The large hydronephrotic kidney is susceptible to rupture by minimal trauma and the hydronephrotic horseshoe kidney far more so. It lies lower down: it is not protected by the ribs and costal margin. Moreover, it lies nearer the middle line and cannot move away when any force is applied to it.

A man of 21 was struck in the abdomen by a football. Laparotomy forty-eight hours later revealed dirty peritoneal fluid arising from a hydronephrotic left kidney. The rupture in the pelvis was repaired, a nephrostomy performed and the peritoneal cavity drained. Subsequent retrograde pyelograms revealed a horseshoe kidney, the left half of which was later removed.

Renal parenchymal disease: The horseshoe is fully liable to the usual kidney diseases: nephritis, tubercle, polycystic disease, solitary cysts and

malignant tumours, either hypernephromata or Wilms's tumours. My series contained no tumours but there was one patient with tubercle and Fig 5 shows a case of solitary cyst.

Leriche syndrome: Horseshoe kidneys are further associated with the Leriche syndrome. In this condition there is a thrombosis of the terminal aorta with claudication of the buttocks and lower extremities, impotence and diminished or absent pulses beyond the aortic bifurcation. The proximal edge of the thrombus lies beneath the isthmus, which sometimes exerts a constricting influence on the aorta. The inferior mesenteric artery may be enormously enlarged or obliterated (Hubbell 1958).

A male patient of 47 in 1956 had the isthmus divided prior to a thromboendarterectomy: the bridge was causing constriction of the aorta (Fig 6). Later both legs were amputated and in 1966 the patient died from extensive arterial disease.

Symptomatology: The common symptoms are those of pain: renal colic or pain around the umbilical region attributable to urinary tract infection. The severe pain is that of an acute distension or hydronephrosis caused by failure of the pelvis to empty. The horseshoe syndrome (Rovsing's syndrome) is characterized by nausea, vomiting and abdominal pain accentuated by hyperextension. This pain is to be attributed to distension of the pelvis rather than, as formerly thought, to stimulation of nerve plexuses on the posterior abdominal wall. The pain may, however, be exceptionally sited in the epigastrium or umbilical region. Rovsing's sign is as follows: when the patient is standing with the back muscles contracted to hyperextend the spine, characteristic pain is experienced which disappears when the patient lies on his back (Gutierrez 1934). I am not convinced of the diagnostic value of this sign and have never used it.

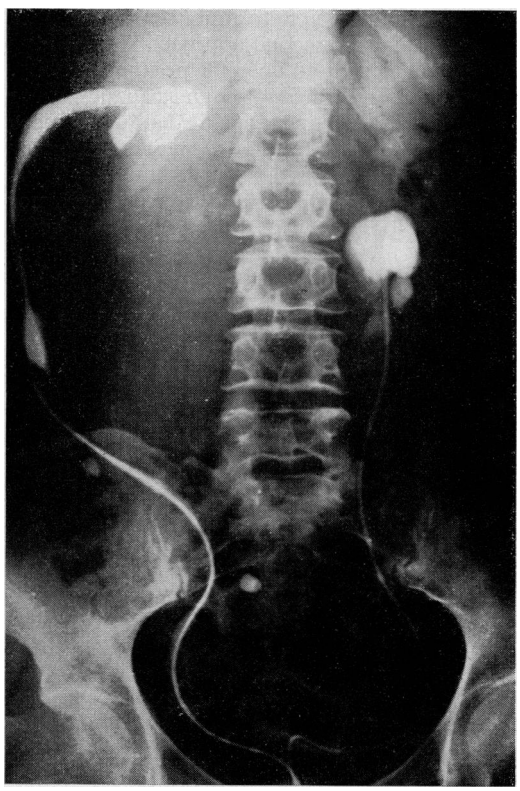

Fig 5 *A solitary cyst in a horseshoe kidney*

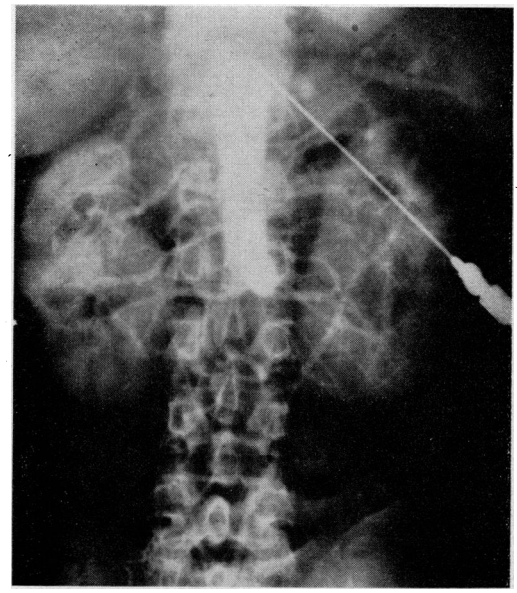

Fig 6 A

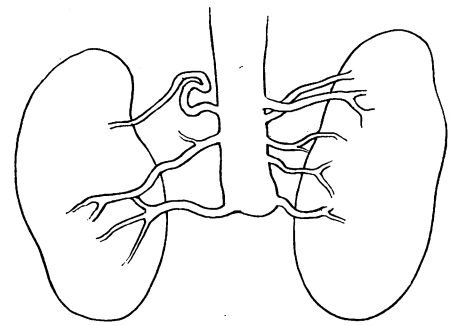

Fig 6 B

Fig 6 *Aortogram (above,* A) *and key diagram (below,* B) *of the patient with the Leriche syndrome*

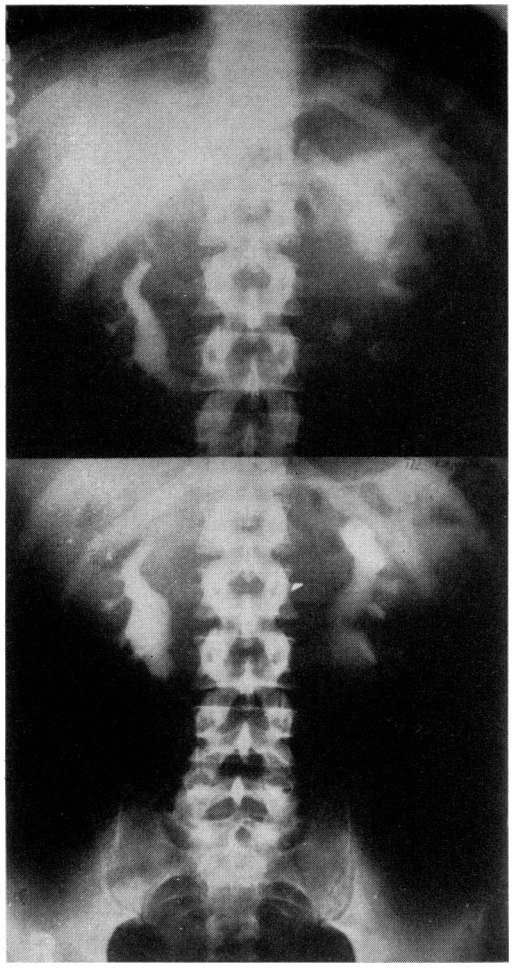

Fig 7 *Pre- and post-operative intravenous urograms*

Fig 7 shows the pre- and post-operative intravenous pyelograms of a male patient of 28, who had left renal colic in 1958, after suffering indefinite pain in the left loin for two years. His symptoms were relieved by division of the isthmus, rotation of the left kidney and a left nephropexy.

Diagnosis: Horseshoe kidneys can seldom be diagnosed by abdominal palpation: they are, however, normally revealed by intravenous urograms. The axes of the kidneys point inferiorly and medially, with calyces projecting medially into the isthmus: the isthmus may be apparent on a straight X-ray. Aortograms and renal arteriograms provide additional information which may be very helpful if surgery is contemplated. The ureters usually lie more medially but this is not necessarily the case.

Treatment: In approximately 25% of cases no treatment is needed; surgery is necessary only when complications supervene. It is indicated in the following conditions: pyelonephritis failing to respond to antibiotics, hydronephrosis, pyonephrosis and stones. Conservative surgery involves symphysiotomy allowing the lower poles to fall apart, nephropexy with rotation and, rarely, excision of the isthmus. With a stricture at the pelvi-ureteric junction or a high origin of the ureter, a plastic operation should be considered on the pelvi-ureteric junction and, with attention to these points, one's failures should be fewer. Nephropexy and rotation of the kidneys may help drainage but the arrangement of the vessels often precludes rotation. Rarely a prominent isthmus should be removed as the upper ureter may become densely adherent to the divided isthmus and produce a secondary obstruction (Fig 8 and C N in Table 1).

Surgery: An adequate exposure is essential. I prefer (*a*) an extraperitoneal approach, an oblique incision running forwards from the anterior end of the eleventh to twelfth rib to near the middle line in the suprapubic region. The operation side of the patient is raised so that the opposite ureter is visible, as would be necessary for a nephrectomy involving division of the isthmus. It is, however, impossible to mobilize the opposite kidney so that the second approach is (*b*) a long middle line incision to open the peritoneal cavity. The cæcum, ascending colon and hepatic flexure are mobilized and retracted medially to expose the fused kidney, the inferior mesenteric artery being identified and avoided. The blood supply is studied and the ureters are identified. The lower group of blood vessels to the kidneys and any

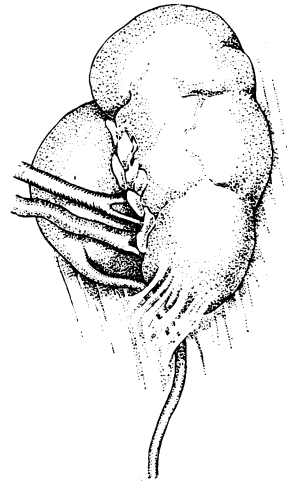

Fig 8 *Secondary obstruction due to adhesions between the upper ureter and the divided isthmus*

Table 1
Cases of horseshoe kidney treated surgically

Case	*Age*	*Symptoms*	*Findings*	*Operation*	*Result*
Hydronephrosis: 5 cases					
P H	28	Left colic	Left hydronephrosis	Symphysiotomy, nephropexy	Cured
G G	14	Right colic	Right hydronephrosis	Symphysiotomy. Secondary nephrectomy	Non-functioning. Nephrectomy
F W	10	Left colic	Normal	Symphysiotomy	Cured
R E D	21	Right colic	Hydronephrosis, right more than left	Symphysiotomy, right nephropexy	Cured
C N	10	Left colic	Bilateral hydronephrosis, left more than right	Symphysiotomy, bilateral nephropexy	Right cured, left worse
				Secondary exploration of left kidney	Improved
Hydronephrosis: 2 cases in solitary kidneys					
J G	22	Right colic and anuria	Right hydronephrosis ●	Plastic right pelvi-ureteric junction nephropexy	Cured
P F	23	Right colic, anuria	Right hydronephrosis. Left, non-functioning	Symphysiotomy	Anuria. Right nephrostomy
Hydronephrosis and infection: 2 cases					
R B	6	Recurrent infection: ? pyelonephritis	Bilateral hydronephrosis	Symphysiotomy and right nephropexy	Improved
F F	59	Left pain	Left pyonephrosis	Left nephrectomy	Cured
Hydronephrosis, infection and stones: 2 cases					
L C	56	Persistent infection	Bilateral hydronephrosis, bilateral stones	Symphysiotomy, left pyeloplasty ■	
D W	2	Infection	Bilateral hydronephrosis, bilateral stones	(1) Symphysiotomy, right nephropexy and ureterolithotomy	Satisfactory
				(2) Left pyelolithotomy and nephropexy	Satisfactory
Miscellaneous cases					
R G	21		Ruptured kidney. Intraperitoneal rupture of enormous left hydronephrosis	Left nephrectomy	Cured
A M	23		Tuberculous right kidney and tuberculous stricture of right ureter	Cutaneous ureterostomy	Improved
R J	47		Horseshoe kidney and Leriche syndrome	Symphysiotomy	Improved

● Left kidney removed five years before for stones
■ Developed hypertension and left renal artery stenosis and had left nephrectomy

Table 2
Cases of horseshoe kidney treated conservatively

Case	*Age*	*Symptoms*	*Findings*	*Additional findings*	*Result*
V Z	11	Infection	No hydronephrosis	–	Infection controlled
C C	67	No infection	No hydronephrosis	Prostatic obstruction	Prostatectomy
K M	41	No infection	No hydronephrosis	Prostatitis	
A K	37	Recurrent infection	No hydronephrosis	–	Infection controlled
W L	16	No infection	No hydronephrosis	Bilateral loin pain	Patient refused further investigation and treatment
H H F	68	No infection	Bilateral hydronephrosis	Prostatic obstruction	Prostatectomy

vessels to the isthmus must be dissected out. Any large vessels to the isthmus will require to be ligated and divided. Before dividing a 'fat isthmus' it is useful to occlude any large artery to the lower half of the kidney with a bull-dog clamp. The vessels in the cut isthmus can then be underrun with fine catgut. The incision in the isthmus is not necessarily in the middle line and care is taken not to open a dilated calyx. Fat is then placed on the raw surface and a nephropexy is performed. Resection of the isthmus is occasionally necessary to allow the ureters to lie medial to the lower pole of the kidney. At the end of the operation a drain is brought out retroperitoneally. Dahlen & Schlumberger (1957) advocated a transperitoneal approach through an avascular area in

the posterior peritoneum directly over the isthmus and this could be combined with my approach. Geoffrey Parker (1956) described an incision 2 in. (5 cm) below and parallel to the costal margin but I have not used it. Robinson (1936) described 4 cases, in one of which a periarterial sympathectomy was performed, a thick cortical isthmus being present; the patient's symptoms were relieved.

Personal series: My cases total 20, the youngest patient being 2 years old and the eldest 68. Only 3 were females. Thirteen patients have been operated on, and another one has had a cutaneous ureterostomy for tubercle. I have not recorded patients who have been seen once only.

The cases fall into the groups shown in Tables 1 and 2.

Of the failures, 2 patients have required nephrectomy, while the third, a case with a solitary kidney, has a permanent nephrostomy, performed abroad. In all 3 cases a symphysiotomy, rotation of the kidney and nephropexy were performed. The question arises whether some form of plastic operation should have been added: further, whether rotation and nephropexy, if feasible are always necessary.

Summary and Conclusions

(1) Twenty-five to thirty per cent of horseshoe kidneys need no surgery and any operation should require cast-iron indications.

(2) The most usual finding is unilateral or bilateral hydronephrosis with all the associated complications.

(3) Operative treatment should be directed to overcome all obstructions.

(4) In a small number of cases the initial operation will be followed by obstruction (possibly not relieved by the operation or a secondary obstruction between upper ureter and the divided isthmus).

(5) Short and long term follow up must be done, short follow up to pick up any increase in hydronephrosis. No long term follow ups are available anywhere: 10 of my operation cases are under 30 and should be seen regularly over the next forty to fifty years.

(6) I agree with what Edmund Papin wrote in the foreword to Gutierrez's monograph (1934) on horseshoe kidney: 'If the horseshoe is regarded by superstitious persons as a source of good luck, this is not the case with horseshoe kidney which is, on the contrary, a lamentable infirmity.'

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