Correspondence

An unusual case of complete renal fusion giving rise to a 'cake' or 'lump' kidney

It is well known that some types of renal abnormalities are relatively commonly encountered; in a number of large studies, it has been estimated that urinary tract anomalies are observed in about 10% of all autopsies (Rubenstein et al. 1961). Similarly, renal tract abnormalities often coexist with anomalies of the cardiovascular, central nervous, genital tract and skeletal system and abnormalities of the gastrointestinal tract, with anorectal anomalies being particularly commonly encountered in these individuals (Boatman et al. 1972; Cook & Stephens, 1977; Van Allen, 1993; Bauer, 1998). Horseshoe kidney, for example, is observed in about 20% of individuals with trisomy 18 and in at least one third of females with Turner's syndrome (Smith & Jones, 1970; Lippe et al. 1988). It is relevant that before the introduction of pyelography, all cases of renal abnormalities were found either at autopsy or operation (Wilmer, 1938).

Possibly the rarest of these renal abnormalities involves the complete fusion of the 2 kidneys to form a single entity sometimes referred to as a 'cake' ('Kuchenniere') or 'lump' kidney. According to Wilmer (1938), Pannorlus first described this condition in 1654 [the name of this author is given as 'Pamarolus' by Bauer (1998)], but no further details are provided of this publication in either source. This condition represents an extreme variant of the 'horseshoe' kidney, a condition first described by da Carpi (1522). The latter condition, accounts for 90% of all renal fusion anomalies and occurs with an incidence of about 0.25% of the population, or about 1 in 400 at postmortem examination (Warkany, 1971; Bauer, 1998), although such kidneys have been found in 1 in 200 individuals on radiography (Van Allen, 1993). In horseshoe kidneys, only the 2 lower poles are normally fused together, although fusion of both the upper and lower poles may occur, resulting in a disk or doughnut shape, an anomaly that accounts for only 2% of these fused kidneys (Warkany, 1971). The ureteric buds do not cross the midline before entering the future renal pelvises. Such kidneys are unable to ascend to their normal upper lumbar location, being impeded by the presence of the inferior mesenteric vessels. As in other renal fusion anomalies, it is found more commonly in males with a male:female ratio of about 2–3:1. Nongenitourinary tract anomalies were present in one-third of 96 individuals with horseshoe kidneys studied by Boatman et al. (1972; for a detailed discussion of the pathological conditions associated with crossed renal ectopia in which the ureters may or may not cross the midline to insert into the opposite side of the bladder, see McDonald & McClennan, 1957; Bauer, 1998). The term 'crossed renal ectopia' has been defined as a 'congenital condition in which a ureter in the normal bladder position crosses the midline to an ectopic kidney lying on the opposite side of the body' (McDonald & McClellan, 1957). When the ectopic kidney is located in the pelvis, the ureters would not normally cross the midline.

In the examples of complete renal fusion described in the literature, such 'cake' or 'lump' kidneys are almost invariably located in the midline and are most commonly observed in the pelvis. With very few exceptions, such kidneys possess 2 ureters, both of which enter the bladder in the normal regions of the trigone (Bauer, 1998). Two rare examples of fused kidneys of the 'lump' variety have been described in which only a single ureter was present (McCrea, 1942; Brock et al. 1983). Clearly, these would have to be distinguished from the considerably more commonly encountered solitary kidney which may or may not be located in the pelvis, and which only possesses a single ureter (Berg & Kearns, 1949). In a number of the reported cases of fused kidneys, this condition was associated with anomalies of the cloacal derivatives and lower limbs in what has become termed the 'caudal regression syndrome' (Duhamel, 1961; Braren & Jones, 1978; Brock et al. 1983).

In all cases of fused kidneys, the arterial supply and venous drainage are grossly abnormal, and this reflects the primitive arrangement invariably seen in ectopic kidneys, often because of their very limited rostral migration. The particularly unusual feature seen in the example described here is that the fused kidney was exclusively located to the right of the midline in the lumbar region. It is of interest that a photograph of an example of a 'lump' kidney located in the right lumbar region similar to that observed in the present case is illustrated by Risdon in Heptinstall's *Pathology of the Kidney* (see fig. 2–22, p. 123, Risdon, 1992).

The majority of cases of fused kidneys described in the literature have been observed at autopsy in patients who had severe renal manifestations (e.g. Wilmer, 1938, in which a total of 286 cases were reviewed; Kron & Meranze, 1949; Shiller & Wiswell, 1957; Glenn, 1958; Srivastava et al. 1971). The most commonly encountered symptom present is pain due to one of 3 possible causes: the anomalous kidney pressing on certain visceral structures, renal lesions, and lesions not associated with the kidney. This is, however, not invariably the case, and the example described here was observed in a cadaver during the normal course of its anatomical examination. The subject was an 81-y-old woman, with no clinical history of renal disease at the time of her death, and many examples of this condition have been described in the literature where there were no symptoms of renal origin.

The 'cake' or 'lump' kidney was located in the right lumbar region. It appeared to be a solitary mass of renal tissue, with no obvious evidence of lobulation, a common feature of such kidneys. The upper pole of this kidney was at about the level of the 3rd lumbar vertebral body, while the caudal pole was just above the level of the sacral promontory. Both ureters originated from the anterior aspect of the fused kidney. The right ureter took origin from a pelvis located close to the medial aspect of the rostral pole

of the kidney. The left ureter, which crossed the midline to enter the left side of the bladder, originated from close to the anterior surface of the central region of the fused renal mass. The 2 ureters appeared to enter the bladder in the normal location. Neither ureter showed any of the features of hydronephrosis, and there was no evidence of vascular compression at any point along their course, or close to their sites of entry into the bladder.

An inspection of the lumbar region revealed that the descending abdominal aorta was located mainly in the midline. The inferior vena cava was located slightly further to the right side than is normally the case in all but the most caudal part of its course where it was dorsal to the bifurcation of the abdominal aorta. The arterial supply of this fused kidney was anomalous, being derived largely from a system of right renal vessels. Apart from the most caudal renal artery the origin of which was located in the midline just rostral to the bifurcation of the abdominal aorta, and which subsequently gave off 2 branches which supplied the majority of the fused renal mass, the 2 other renal arteries present were both derived from the right side of the aorta. The most rostral renal artery present was of narrower diameter and supplied the lateral side of its rostral pole. The third renal artery present had a similar diameter to the caudal renal artery and was located approximately midway between the origin of the other 2 vessels. It principally supplied the medial part of the upper pole of the kidney, close to the right renal pelvis.

The venous drainage was via a number of relatively wide diameter vessels the most rostral of which were 2 veins that appeared to drain the upper pole of the kidney. One of these vessels drained into the right side of the inferior vena cava, while the other drained into the ventral part of the inferior vena cava at about the same vertebral level. The majority of the kidney, however, drained via 2 branches that joined to form a single vessel arising from the most caudal part of the inferior vena cava. The origin of this vessel was just rostral and slightly to the left side of the confluence of the 2 common iliac veins and was just caudal and deep to that of the caudal renal artery (see above). Along the majority of their length, the course of the 2 renal veins was directly anterior to the corresponding renal arteries. Just distal to the confluence of these 2 veins, the single draining vessel coursed caudally and ran behind and deep to the corresponding artery before joining the inferior vena cava. The principal anatomical features of this fused kidney and its anomalous vascular arrangement are shown in the Figure.

The left adrenal vein was joined at its midpoint by the left ovarian vein, and drained into the left side of the inferior vena cava about 2 cm rostral to the renal vein that drained the upper pole of the kidney. The right adrenal vein entered the right side of the inferior vena cava about 2 cm rostral to the level of entry of the left adrenal vein. This vessel is not displayed in Figure 1. No right ovarian vein was noted.

Access to the departmental records of this individual revealed that she had had children, and that the absence of her uterus was due to a hysterectomy, although it is unclear whether one or both ovaries were removed at that time. In the absence of access to her clinical records it was not possible to ascertain whether the right ovary was ever present, in the light of the absence of the right ovarian vein (see above). The presence of a left ovarian vein but absent left ovary strongly suggests that the left ovary had probably been removed when the hysterectomy was performed. The only other abdominal feature observed was evidence of a partial gastrectomy that appeared to have been carried out many years earlier. No obvious anorectal abnormalities were observed. As a radiological examination of the trunk and pelvic region was not performed, it was not possible to establish with any degree of certainty whether there were any associated vertebral abnormalities.

While the first reported case of a completely fused pelvic kidney in the modern literature dates from Huntington's Harveian Society Lecture of 1907 (Huntington, 1908), it is generally recognised that the first *detailed* report was that of Looney & Dodd (1926). By 1957, it was believed that only 9 examples of this condition had previously been reported (Shiller & Wiswell, 1957), although some doubt was expressed at that time regarding the anatomical features of some of the earlier examples. A number of attempts have subsequently been made to establish the frequency of fused pelvic kidneys, and in one study 3 cases were observed in 51 800 consecutive autopsies, giving an incidence of about 1 in 17 250 cases (Campbell & Harrison, 1970). In an earlier review of the literature, Wilmer (1938) indicated that unilateral fused kidneys were observed in approximately 1 per 7500 autopsies, although if the figures from Rush Medical College were excluded from this analysis, the incidence was closer to 1 per 12 000 cases.

Bauer (1998), in his account of crossed ectopic kidneys with fusion, published in a subsequent edition of Campbell's *Urology*, was only prepared to state that 'the lump or cake kidney is a relatively rare form of fusion. … The total kidney mass is irregular and lobulated. Generally, ascent progresses as far as the sacral promontory, but in many instances the kidney remains within the true pelvis. Both renal pelves are anterior and drain separate areas of parenchyma. The ureters [characteristically] do not cross.' This author distinguished the 'lump' type of fused kidney from the following types: inferior ectopic kidney; sigmoid or S-shaped kidney; L-shaped (also termed tandem) kidney; disc (also termed shield, doughnut or pancake) kidney, and the superior ectopic kidney. The 2 most unusual variants reported, those with a single ureter, and the type represented by the present example, which appears to have 'ascended' to the midlumbar region, are therefore likely to be even less commonly encountered.

Few suggestions are available in the literature which help to explain the early development of the 'lump' kidney. Srivastava et al. (1971), have proposed that 'at the 9 mm stage (about 30 d of gestation), the nephrogenic blastemas are squeezed together by the umbilical arteries,' and that this may cause their fusion. They continued 'complete renal fusion may be explained by an abnormal course of the ureteric buds, fusion occurring if their terminations are too closely approximated. Alternatively, this anomaly could result from the growth of the ureteric buds into a common metanephric blastema.' We are of the view that this latter suggestion is the most likely possibility. This explanation is consistent with that provided by Cook & Stephens (1977), who suggested that the 'development of a fused kidney [forms] from a single nephrogenic cord which has been met by the ureteric buds arising from both wolffian ducts.' They further suggested that 'the position of the ureteric buds

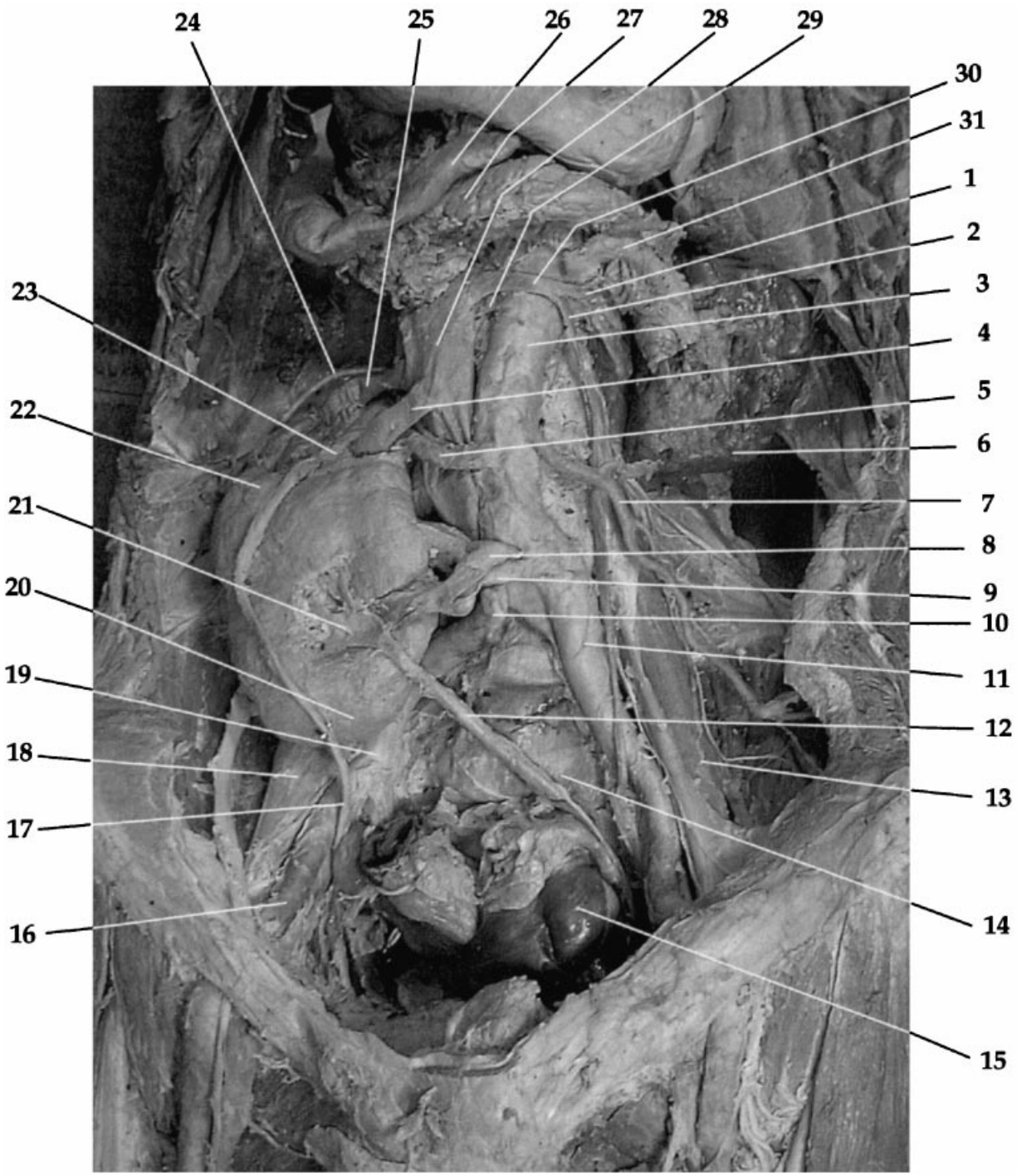

Fig. 1. Gross appearance of the abdomen dissected to display the presence of a 'cake' or 'lump' kidney in situ. 1, Left adrenal vein; 2, distal part of left ovarian vein; 3, abdominal aorta; 4, rostral renal vein draining upper pole of fused kidney; 5, rostral renal artery; 6, spleen; 7, inferior mesenteric artery; 8, principal renal vein draining the majority of the fused renal mass; 9, principal renal artery supplying the majority of the fused renal mass; 10, right common iliac artery; 11, left common iliac artery; 12, left ureter; 13, left psoas major muscle; 14, sacral promontory; 15, sigmoid colon; 16, right external iliac artery; 17, right ureter; 18, right psoas major muscle; 19, bifurcation of right common iliac artery; 20, lower pole of fused kidney; 21, left renal pelvis; 22, upper pole of fused kidney; 23, right renal pelvis; 24, small diameter renal artery passing to upper pole of kidney; 25, additional renal vein draining the upper pole of the fused kidney; 26, terminal part of third part of the duodenum (displaced rostrally); 27, body of pancreas; 28, inferior vena cava; 29, origin of most rostral renal artery passing to the upper pole of the fused kidney; 30, vein draining left ovarian and left adrenal veins to inferior vena cava, 31; upper pole of left adrenal gland.

relative to the midline may be affected by abnormal orientation [for example, lateral flexion] of the 'tail' relative to the trunk.' It must be assumed that the nephrogenic tissue from the side that does not receive a ureteric bud not only fails to develop into a definitive kidney, but completely regresses.

Other authors have also concluded that the position, fetal lobulation, and pelviocalyceal configuration indicate that development is arrested at the 10 mm stage (Glenn, 1958; Brock et al. 1983). Such kidneys may show the following histological features: immature glomeruli, cystic changes (which may be haemorrhagic) and enlarged dilated tubules (Shiller & Wiswell, 1957), or evidence of long-standing renal disease (Kron & Meranze, 1949). In other cases, there may be evidence of infarction secondary to the abnormal blood supply (Saylor & Gordon, 1951). In the case described by Saylor & Gordon (1951), renal infarction was the immediate cause of death. The situation was complicated by aortic thrombosis with occlusion.

While increased numbers of cases of fused kidneys are now being recognised in clinical practice with ultrasound, intravenous pyelography and CT scanning, few examples are seen by anatomists in the normal course of their activities. This is likely to be because of the rarity of this condition and the fact that a proportion of individuals present with renal complications or may die due to complications resulting from the presence of associated congenital abnormalities. Surgical correction of, for example, obstructed and hydronephrotic ureters, which may lead to the development of urinary tract infection and renal calculi, which occur more commonly in fused kidneys than in horseshoe kidneys, tends to be extremely difficult not only because of their aberrant blood supply but because of the unusual structure of the pelves and ureters (Bauer, 1998). Having listed the clinical complications that may be associated with fused kidneys, Bauer (1998) has recently emphasised that most individuals with this condition have a normal longevity and prognosis, as appears to have been the case with our subject.

The fact that this individual had had children before a hysterectomy had been performed was of interest, because congenital absence of the vagina and absence or abnormalities of the internal genital tract may occasionally be associated with this condition (Cook & Stephens, 1977; Risdon, 1977). As it was decided that this example represented such a rare abnormality of the renal system, the decision was taken that no histological or further anatomical examination would be performed in this case.

M. H. KAUFMAN[1]* AND G. S. FINDLATER[1]
Section of Anatomy, *Department of Biomedical Sciences*, *Hugh Robson Building*, *George Square*, *Edinburgh EHS 9XD*, *UK*Tel: +44 (0) 131 650 3113; fax: +44 (0) 131 650 6545; e-mail: *M. Kaufman*@ed.ac.uk
* Author for correspondence.

REFERENCES

BAUER SB (1998) Anomalies of the kidney and ureteropelvic junction. In *Campbell's Urology*, (ed. Walsh PC, Retik AB, Vaughan ED, Wein AJ), 7th edn, vol. 2, pp. 1708–1755. Philadelphia: W. B. Saunders.

BERG OC, KEARNS WM (1949) Solitary pelvic kidney: a case report. *Journal of Urology* **62**, 275–277.

BOATMAN DL, KÖLLN CP, FLOCKS RH (1972) Congenital anomalies associated with horseshoe kidney. *Journal of Urology* **107**, 205–207.

BRAREN V, JONES WB (1978) Sacral agenesis: diagnosis, treatment and followup of urological complications. *Journal of Urology* **121**, 543–544.

BROCK JW, BRAREN V, PHILLIPS K, WINFIELD AC (1983) Caudal regression with cake kidney and a single ureter: a case report. *Journal of Urology* **130**, 535–536.

CAMPBELL MF, HARRISON JH (1970) *Urology*. 3rd edn, p. 1447. Philadelphia: W. B. Saunders (cited by M. I. Van Allen).

DA CARPI JB (1522) *Isogogae breves*. '*A Short Introduction to Anatomy*.' (transl. Lind LR). Chicago: University of Chicago Press. 1959 (cited by Cook and Stephens).

COOK WA, STEPHENS FD (1977) Fused kidneys: morphologic study and theory of embryogenesis. *Birth Defects* **13**, 327–340.

DUHAMEL B (1961) From the mermaid to anal imperforation: the syndrome of caudal regression. *Archives of Disease in Childhood* **36**, 152–155.

GLENN JF (1958) Fused pelvic kidney. *Journal of Urology* **80**, 7–9.

HUNTINGTON GS (1908) The genetic interpretation and surgical significance of some variations of the genito-urinary tract. The Harvey Lectures, 1907–08, pp. 222–267. Philadelphia and London: J. B. Lippincott (cited by Shiller and Wiswell: according to Wilmer, this refers to the Harvey Lectures of 1906–07).

KRON SD, MERANZE DR (1949) Completely fused pelvic kidney. *Journal of Urology* **62**, 278–285.

LIPPE B, GEFFNER ME, DIETRICH RB, BOECHAT MI, KANGARLOO H (1988) Renal malformations in patients with Turner syndrome: imaging in 141 patients. *Pediatrics* **82**, 852–856.

LOONEY WW, DODD DL (1926) An ectopic (pelvic) completely fused (cake) kidney associated with various anomalies of the abdominal viscera. *Annals of Surgery* **84**, 522–524.

McDONALD JH, McCLENNAN DS (1957) Crossed renal ectopia. *American Journal of Surgery* **93**, 995–1002.

McCREA LE (1942) Congenital solitary pelvic kidney. *Journal of Urology* **48**, 58–68.

RISDON RA (1977) Development, developmental defects, and cystic diseases of the kidney. In *R. H. Heptinstall's Pathology of the Kidney*, 4th edn, pp. 93–167. Boston: Little, Brown.

RUBENSTEIN M, MEYER R, BERNSTEIN J (1961) Congenital abnormalities of the urinary system. *Journal of Pediatrics* **58**, 356–366.

SAYLOR M, GORDON H (1951) Crossed renal ectopia with fusion complicated by massive infarction due to occlusive thrombosis of the aorta, a case report. *Journal of Urology* **65**, 968–971.

SHILLER WR, WISWELL OB (1957) A fused pelvic (cake) kidney. *Journal of Urology* **78**, 9–16.

SMITH DW, JONES KL (1970) *Recognizable Patterns of Human Malformation. Genetic, Embryologic and Clinical Aspects*. 3rd edn. Philadelphia: W.B. Saunders.

SRIVASTAVA RN, SINGH M, GHAI OP, SETHI U (1971) Complete renal fusion ("cake"/"lump" kidney). *British Journal of Urology* **43**, 391–394.

VAN ALLEN MI (1993) Urinary tract. In *Human Malformations and Related Anomalies*. (ed. Stevenson RE, Hall JG, Goodman RM), Volume 2. New York and Oxford: Oxford University Press, 501–550.

WARKANY J (1971) *Congenital Malformations. Notes and Comments*, pp. 1050–1053. Chicago: Year Book Medical Publishers.

WILMER HA (1938) Unilateral fused kidney: a report of five cases and a review of the literature. *Journal of Urology* **40**, 551–571.