Multicystic Kidney Disease in Children

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I am pleased and honoured to have the opportunity to write this article in the volume which is honouring the memory of the late Herbert B. Eckstein.

I knew Herbert from the time when we were both on the house at Gt. Ormond Street Children's Hospital, until his sad death in 1986. He had by then already made his mark in Pediatric Urology in Turkey, having done such splendid work there on calculus disease in children. He was later to increase his fields of interest to include all aspects of both Pediatric Surgery and Urology including his special interest in Spina Bifida and all its manifestations.

He was an avid reader, a good pediatrician and an excellent surgeon. He was able to set his mind and effort to any task which confronted him. He was also a good lecturer - and was most popular in Europe where his ability to speak in languages other than English made him especially sought after. He wrote extensively on all aspects of his chosen field, and together with R. Hohenfellner and D.I. Williams edited the excellent book on Surgical Pediatric Urology [1]. His only unwavering obsession was the use of the anterior intraperitoneal approach for surgery of the pelviureteric junction. This he preached in both lectures and articles, in spite of the many attempts made by his colleagues to convince him that the antero-lateral extraperitoneal approach was far superior. [*]

It is my privilege to write this chapter on:

Multicystic kidney disease in childhood (MCKD)

A great deal of confusion has surrounded the topic of cystic disease and much of this can be accounted for by the loose usage of the prefixes Poly- and Multi. In addition any dilatation or even any abnormal saccular area is often referred to as a cyst. Clarification and precise terminology must be established before any attempt is made to discuss the topic.

The Report of the Committee on Classification, Nomenclature and Terminology, Section of Urology of the American Academy of Pediatrics by K. I. Glassberg, et al [2] goes a long way to elucidating this problem. They discuss many aspects of renal development which may be associated with or responsible for the condition of Cystic Kidneys, suggest precise terminology and outline a classification of renal cystic disease.

I will refer to some of the terms and definitions which they recommend:

Dysplasia is due to abnormal metanephric differentiation - and it is diagnosed histologically- the only irrefutable evidence being the presence of primitive ducts and nests of metaplastic cartilage. Cyst may or may not be present.

Aplasia is an extreme form of dysplasia with only a nubbin of tissue remaining.

Atrophy is loss of parenchyma due to known or unknown causes.

Hypoplasia refers to a small kidney or kidney segment.

Reflux nephropathy - a label for any abnormal renal morphology associated with vesico-ureteric reflux.

They suggest that Cystic Disease of the Kidneys can conveniently be divided into two main categories (Table 1).

This article will only discuss the topic of MCKD. However, for a sound understanding of the subject, one should have a working knowledge of all the causes of cystic kidney disease listed in Table 1, and be especially conversant with the genetic as opposed to the non genetic forms.

History

Cruveilhier [3] in *Anatomie Pathologique du Corps Humain* (1836) demonstrated in an autopsy

Table 1. Cystic disease of the kidneys

Genetic:	
	Infantile (autosomal recessive polycystic kidneys)
	Adult (autosomal dominant polycystic kidneys)
	Juvenile nephronophthisis-Medullary cystic disease complex
	Juvenile nephronophthisis (autosomal recessive)
	Medullary cystic disease (autosomal dominant)
	Congenital nephrosis (autosomal recessive)
	Cysts associated with multiple malformation syndromes
Non genetic:	
	Multicystic kidney (Multicystic dysplasia)
	Multilocular cysts (multilocular cystic nephroma)
	Simple cysts
	Medullary sponge kidneys (less than 5% inherited)
	Acquired renal cystic disease-in chronic hemodialysis
	Caliceal diverticulum (pyelogenic cyst)

specimen of a 3 year old boy, a kidney which contained multiple cysts but with no trace of normal parenchyma; his drawing of the specimen shows it to be what we now class as MCKD.

The first true understanding of the entity, and its separation from other cystic lesions of the kidneys was made by Spence in 1955 [4]. He reviewed the literature and described four additional cases. His cases were perhaps unusual as two of the four were in adults and they were symptomatic. Nevertheless he was the first to name the condition MCKD and to differentiate it from the other cystic lesions of the kidney.

Many cases and reviews have appeared since, but is has been the advent of sonography (US) that has changed the whole concept of the problem. The serendipitous discovery of fetal renal pathology by prenatal sonography has changed our concepts, not only of MCKD, but also of many other detectable renal lesions.

Sex and side

Males seem to be affected slightly more frequently than females and the left side more than the right. Very occasionally the condition is bilateral but is then incompatible with life.

Symptomatology

Abdominal mass: The discovery of an asymptomatic abdominal mass, either visible or palpable by the parents or physician and accompanied by the finding of a non-functioning kidney on the same side, used to be the commonest clinical method of presentation in infancy and childhood. As more than 75% of the abdominal masses found were genito-urinary in origin, and more than 60% were due to MCKD, the condition was fairly readily diagnosed.

Today the use of US to evaluate the state of the pregnancy (for its duration, the number of fetuses and their condition etc.,) has changed all that, for the finding of cysts within the kidney area and little or no kidney cortex points to the possible diagnosis of MCKD. Prenatally an increased number of cases are being suspected of being MCKD, and are confirmed postnatally, but many of these are found to be clinically asymptomatic and have No Palpable Mass. They would therefore have been missed were it not for the prenatal US screening. This investigation alone has increased the numbers of cases that are being diagnosed.

Urinary tract infection and hematuria:

Previously when a patient with UTI was being investigated; a mass and a non-functioning kidney were occasionally found, leading to the suspected diagnosis of a MCKD. It was assumed that those symptoms were arising from the MCKD, but this may not have been so. Further investigation of these cases demonstrated that the symptoms could well have originated in the opposite kidney or ureter which are quite often affected by hydronephrosis or reflux. This has been shown to be so in up to 50% of cases of MCKD [5,6]. This leads to the vexed questions as to whether multicystic kidneys can ever be the seat of infection. Early on it was thought that this was possible and very rarely cases were reported where a removed multicystic kidney was surrounded by inflammatory exudate.

Let us also consider if it is at all possible that the involved kidney can be the seat of the urinary infection taking into account its pathology. Firstly there is often atresia or stenosis of its ureter thus eliminating the possibility of the infection descending into the bladder. Secondly there is rarely a pelvis on that side and little if any recognisable renal elements. Thus how can infection that is supposed to arise from the MCKD reach the bladder if these abnormalities are present?

Taking into account all the criteria listed above it seems highly unlikely, as in most cases the infection probably arises from the opposite kidney or ureter or theoretically even from its own non communicating but abnormal lower ureter.

Hypertension: There have been a few possible cases of MCKD associated with or presenting as hypertension. Susskind et al [7] and Gordon et al, [8] in an intensive 20 year literature survey, found 9 cases, but in only 3 of the 9, did surgical removal improve the hypertension. Wacksman and Phipps [9] in a report from the Multicytic Kidney Register found no cases of hypertension in the 441 cases in that series. In Manchester, J Bruce et al [10] of St Mary's Hospital report that they have had 3 cases in the past few years. Notwithstanding, this symptom is extremely rare.

Malignancy: This too is a rarity and is usually associated with, rather than a presenting feature. In the same survey Gordon et al [8], have discovered only 6 cases over the past 20 years - 3 in adults and 3 in children. Their figures and extrapolations hypothesise that there is approximately a 1/3333 chance that a malignancy will occur in MCKD. Nevertheless, because it is undoubtedly of great concern and worry to the parents and physicians, and even though the possibility is so remote, it is the most compelling reason for considering nephrectomy.

Differential diagnosis of the abdominal mass

In the pre-ultrasound era, with the more limited investigatory methods available, differentiating MCKD from other renal pathology was rather haphazard. Today investigation is much more satisfactory and a rapid and accurate diagnosis can readily be made. The differential diagnosis (then as well as now) is mainly between MCKD and hydronephrosis, renal tumours and other cystic renal lesions.

It also has to be differentiated from extra-renal lesions as may occur in the bowel, liver or genital systems, but this is very easily accomplished by US.

Renal lesions

1) Hydronephrosis is the most common lesion that has to be differentiated from the multicystic kidney.

a) Straight x-ray of the abdomen in both lesions may show an abdominal mass with displacement of the bowel if the lesion is large enough. In rare instances there may be calcification in the multicystic kidney (in older patients and notably in adults).

b) The ultrasound picture can often be diagnostic for it shows cysts of varying size, no pelvis, no cortical tissue and no ureter, whereas in hydronephrosis the pelvis is grossly dilated with communication with largely dilated calyces. There is usually some remaining renal tissue except in the most severe cases. In MCKD the opposite kidney is usually normal, but it may show signs of hydronephrosis or of reflux nephropathy.

c) Intravenous urography will show no evidence of function in almost all cases of multicystic kidneys. In very rare cases a small area of puddled contrast may be visible (once again mainly in adults). In hydronephrosis however the kidney will usually demonstrate varying degrees of concentration of the dye depending on the severity of, and the damage to the kidney caused by the pathology. This is more pronounced in a dilated pelvis and calyces.

d) Retrograde urography can be used to differentiate between the two. In hydronephrosis the dilated pelvis and calyces are confirmed. The ureter is in continuity, but has an area of relative stenosis of the pelviureteric junction. In contrast in MCKD it is usual to find an atretic or otherwise abnormal ureter with no communication to the cystic area and the pelvis is usually absent.

c) Micturating cystography is useful to demonstrate vesico-ureteric reflux which occasionally may be present on the opposite and even into the atretic or abnormal ureter on the MCKD side.

d) Renography will show a total lack of function on the affected side.

e) Only in very rare instances is the use of Computer Assisted Tomography or Magnetic Resonance Imaging necessary.

2) Renal malignancy (Wilms' tumour) can usually be demonstrated by the above investigatory procedures. It must be noted that occasionally in MCKD there may be an associated malignancy, but these tend to be in the type where more solid elements are present

Natural history

Prior to the advent of ultrasonography the natural history MCKD was never very clearly known or understood. This is because the MCKD was usually discovered in infancy or childhood and was followed by surgical excision, for the reasons mentioned above. Today the majority of cases are being diagnosed prenatally and when followed postnatally are found to be entirely asymptomatic. This allows one time to watch the patient and to evaluate the long term prognosis. Thus in time we will be able to know the true natural history of MCKD.

Treatment

The conservative approach has been adopted in many centres and the results which are now being published are very encouraging.

The consensus is that the majority of cases do not need excision. When followed carefully with regular abdominal examinations and repeated US investigations, the lesions are seen to diminish in size and in some cases even disappear completely. If however any of the following occur, then possible surgical removal may be necessary:

a) Increase in the size of the MCKD being such that it is causing problems by pressure on adjacent structures;

b) Investigations being non-conclusive, and the remote possibility of there being a malignancy may force one to operate;

c) Pressure by the parents, who despite being given all the facts and possibilities, are still overanxious and would prefer to have the mass removed.

In certain circumstances where a large MCKD is causing pressure and especially if there is respiratory embarrassment, it may be expedient to perform a percutaneous drainage reduction of the cystic mass.

Notwithstanding all the above facts, one must still remain aware that there is the possibility that in later years symptoms may appear, hypertension may ensue or even that malignancy may supervene. This has been shown by the reports of cases seen in adults [4,11].

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(*) Editor's Note:
HB Eckstein, "Anterior Approach", in Surgical Pediatric Urology, H.B. Eckstein, R. Hohenfellner, DI Williams (eds), Stuttgart, 1977, p. 102

"The anterior approach to the kidney is remarkably simple and provides an excellent anatomical exposure with minimal dissection. It has attracted relatively little interest in recent years but is advocated as the incision of choice for pyeloplasty, partial nephrectomy, or other surgical procedures on the renal pelvis or renal vasculature. The incision is not recommended for use in calculous disease or in the presence of renal or pararenal infection.

The skin incision may be either transverse or parallel to the costal margin. The transverse incision results in a better scar which may well become virtually invisible in due course, while the Kocher type incision gives a better, more direct approach to the renal pedicle and is especially indicated in children

with an acute costal angle. The incision is deepened by diathermy through the rectus sheath and rectus muscle as well as the external and internal oblique muscles. The transversus abdominis muscle is divided in the line of fibres.

At this stage it is possible either to open the peritoneum extensively in the line of the incision and use a transperitoneal aproach or to carefully dissect off the peritoneum from the abdominal wall musculature and use an extraperitoneal approach. It is important in the latter method to start the separation off the peritoneum from the abdominal musculature at the lateral end of the incision and to dissect forward. In the extraperitoneal approach the renal fascia is displayed and then incised so that the kidney, the renal pelvis and the renal pedicle come into view.

If the transperitoneal approach is used, the intestine is displaced medially and the ascending or descending colon elevated. The peritoneal fold joining the ascending or descending colon to the posterior peritoneum is incised; this will display the renal capsule, which is incised similarly. The colon and the remainder of the intestine are easily retracted by a wet pack with a Dever type retractor and an excellent exposure of the renal pelvis can be obtained with minimal dissecton."

v. also, HB Eckstein and I Kamal: Hydronephrosis due to pelvi-ureteric obstruction in children. An assessment of the anterior transperitoneal approach, Brit J Surg, 58:663-667, 1971