CONGENITAL CYSTIC KIDNEY.

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THIS is the name most frequently given to the condition of which I show a specimen to-night. The term polycystic kidney is more correct, as the congenital nature of the disease is by no means universally accepted.

Cystic kidneys of this sort are sufficiently uncommon to be of interest, being found on an average once in 700 autopsies. The disease presents such a striking appearance that it can never be passed over at a *post-mortem* examination, even though the condition may not have been suspected during life.

Many specimens have been shown at this Section, and we are most of us quite familiar with the condition, this being from the eighth patient I have seen with this disease. In spite of a great deal of work done in connection with polycystic disease of the kidney we know very little about it.

Let me briefly recount the facts that are known in so far as I am familiar with them, then some of the theories advanced, and let us see how they fit in with the rather vague clinical features of such cases.

The disease occurs, or at least manifests itself, at two periods of life—at birth, and between forty and sixty years of age. Isolated cases are found at any age, but the

great bulk occur at the two periods I mention. Out of over 400 cases recorded, about 60 per cent. were found between the ages of forty and sixty.

It is difficult to believe that a disease which develops so late in life is congenital—it is not difficult to suppose that the form seen at birth or soon after is congenital. Are the two the same disease?

I am not familiar with the infantile form, but I believe macroscopically the kidneys are alike in this and in the form occurring late in life. The microscopic appearances vary so much that they are not easy to compare.

There are, however, some points of difference between the two. In the infantile cases, cysts occur more frequently in other organs, chiefly in the liver, and other congenital defects are often found. They do not occur in one sex more than the other. If the child is not born dead, death soon occurs.

In the later forms cysts in other organs occur much less commonly. They have been found, however, in the liver, ovary, testicle, uterus, broad ligament, bladder and the choroid plexus. The disease is much more common in women than in men. Evidence of other congenital defects is rare. The patient may live a long time with this disease.

The polycystic kidney presents a very typical appearance. At first the cysts are mainly on the outer surface of the organ; they vary in colour from white to dark purple, or from yellow to green. They contain a glycerine-like fluid, with numerous epithelial cells. The fluid is never urine, but may contain almost any of its constituents, as urea, calcium oxalate, uric acid, albumin, phosphates, &c. The cystic growth affects the whole kidney, so as the organ enlarges it preserves to some extent its shape. The cysts seem to occur in the con-

volute tubules and are lined with a cylindrical epithelium which in places shows signs of proliferation. Healthy renal tubules may lie between the cysts, but more often the renal tissue left shows signs of chronic interstitial changes. Other changes may occur :—

Hæmorrhage into the cysts.

Septic infection of one or more of the cysts occurs in about 10 per cent. of all cases.

In a few cases *tuberculous infection* has been present.

In advanced cases the disease is nearly always bilateral, but more advanced on one side than the other.

When we catheterise the ureter of such a kidney we get a pale urine of low specific gravity, but otherwise it presents nothing abnormal; only in one-third of the cases is albumin present.

The heart is commonly hypertrophied, the pulse increased in tension.

The disease often affects two or more members of the same family.

Such are the more important facts. What are the theories of origin?

1. That the disease is a new growth.

This theory was first advanced by Malassez, and is held by a host of others who compare the growth to an ovarian cystoma. Some, like Ritchie, look upon the foetal form as congenital and quite separate from the adult form which is believed to be an adenoma.

2. That the disease is inflammatory.

Virchow held this view, that the cysts were really retention cysts, resulting from the gradual blocking of the urinary tubules, as the result of contracting fibrous tissue.

Unfortunately, inflammation is almost never found in the adult form, in the early stages of polycystic kidney, and but rarely in the infantile form. Again, the cystic

kidney that results from extensive interstitial fibrosis is not in the least like the polycystic kidney.

3. The third theory is that of Koster, who believes that all the cases are congenital. Huber has shown that the urinary tubule is developed from two separate structures, a renal vesicle and a collecting tubule, which eventually unite to form a single canal. If this union be imperfect, it is not difficult to understand the cyst formation to be of congenital origin. It is difficult to fit in with this theory the long period of life during which the disease is rare. Busse endeavours to explain this by suggesting that in the adult form the disturbance in development is slight, while in the infantile form it is of a much higher grade.

4. Lastly, a combined theory has been suggested—that the condition in the first instance is an embryonic malformation, and in this a cystomatous growth occurs.

To me none of these theories are satisfactory. The manner in which the disease occurs in several members of the same family, the great frequency with which it occurs in both kidneys—the remarkable similarity of all the specimens, are to my mind all strongly in favour of its being embryonic in origin. What the factor or factors are that prevent the disease developing for the first thirty years of life, and then allow of its rapid development, I do not know, and I have no suggestions to offer on this point.

A word in conclusion about the clinical features. The disease may be divided into three stages—

Stage I.—The cysts are developing in one or both kidneys, but the patient suffers no inconvenience and presents no symptoms.

Stage II.—Pain appears from pressure on surrounding organs, and symptoms of chronic Bright's disease come on.

Stage III.—Uræmic symptoms ending in death.

It is impossible to say how long the first stage may last. Diagnosis only becomes possible when the kidney has attained a fair size. After diagnosis has been made patients have lived as long as twelve years.

The second stage seldom lasts longer than two years, often less. The third stage may last from a few hours to possibly a month or two.

The disease has been responsible for sudden death—that is, death coming on in an hour or two from the first symptom, in persons apparently healthy.

Polycystic kidneys are sometimes only found at an autopsy on such a case.

Acting on the assumption that the disease is a new growth, polycystic kidneys have been removed in the hope of checking the spread of the disease to the second kidney, but in many cases these hopes have not been realised. I know of one lady, well five years after such an operation, but I have since lost sight of her. Seven years is the longest spell of life recorded after operation.

It is noticeable that patients with this disease take a general anæsthetic as a rule very badly.

Stromberg, who has done a good deal of operative work, gives four indications for operation :—

1. Profuse bleeding from the kidney.
2. Pus forming in the kidney.
3. Tumour becoming so large that it causes much pain or dangerous pressure on other organs.
4. Rupture of one or more cysts into the peritoneum.

The removal of such kidneys by operation will give the pathologist opportunities of examining the tumour in its earlier stages, and this combined with more careful examinations of urine obtained by means of the ureteral catheter, may in time throw light on what is still a very obscure subject.

For much of the information in this paper I am indebted to Watson's work on the kidney.

DR. O'KELLY said Mr. Gunn had mentioned that, according to Virchow, cystic disease of the kidney was the result of fibrotic change, but that this change was usually absent when the kidneys were examined. In this connection two cases which recently came under his notice were of interest. In these the kidneys contained many large cysts, and the lesion of chronic interstitial nephritis was present. The cysts were much larger than those usually found in "granular" kidneys, but not so numerous as in cases of cystic disease. The difficulty, which he had not solved, was whether the kidneys were to be regarded as cystic kidneys with chronic interstitial nephritis or as granular kidneys with unusually large cysts. Both kidneys were involved in each case. There was no cystic change in the liver.

DR. BOXWELL was struck by the comparative rarity of cystic kidneys; judging from Mr. Gunn's statistics "one in seven hundred autopsies." Congenital cysts had been mentioned and inflammatory retention cysts. But there was a much commoner form, where the cysts were single or multiple, often very large, and thin-walled, but where there was no evidence of inflammation. These kidneys were continually turning up at autopsies. Were these in an early stage of the condition shown by Mr. Gunn? He had recently seen a very similar change in a sheep's kidney.

PROFESSOR A. H. WHITE said he could not look upon this cyst-formation as a tumour growth at all. He thought it was due either to congenital malformation or to a combination of this with subsequent chronic inflammation and degeneration.

MR. GUNN, in reply, said that perhaps he should have mentioned that the kidney he had shown was taken from a woman about thirty-six years of age, and there was no albumen in the urine before operation. The second kidney had appeared to be normal to the touch. Two days after

the operation the urine was apparently quite normal, and he thought interstitial nephritis was very rarely confined to one side, so that if the kidney which had been removed showed signs of inflammation the second kidney would not be secreting absolutely normal urine. Dr. Boxwell had asked him whether the large cysts one saw in kidneys were the same kind as those met with in the very earliest stage of cystic kidney. It was a multiple disease, and in some cases, where the patient had died, they had found a number of cysts scattered through the kidneys, one kidney being extensively diseased and the other only slightly. It was suggestive that so many cases occurred in women, and between forty and sixty years of age, about the time when the menopause came on.