MEDICAL PRACTICE

Clinical Topics

Clinical aspects of polycystic disease of the kidneys

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Summary

Seventy-eight patients were treated for polycystic disease of the kidneys. An analysis of the interval between the onset of symptoms and end-stage renal failure made it possible to give an accurate prognosis in individual cases. Pregnancy and urinary infection did not appear to accelerate deterioration of renal function, but Rovsing's operation precipitated renal failure in some cases. Forty-two patients needed replacement treatment for end-stage renal failure, and 24 patients received 29 renal allografts. Transplant function at all times was better than a matched group of 70 patients with chronic renal failure from other causes. The only indications for removal of polycystic kidneys in graft recipients were persistent or recurrent infection, erythraemia that failed to respond to conservative treatment, and to make room for the transplant.

Introduction

Cumming described the familial nature of polycystic kidney disease in 1928,[1] and the natural history has been described by several authors.[2-5] Seventy-five per cent of cases are not detected or recognised in life, and until recently those with renal failure died of their disease. Survival has improved since the introduction of dialysis and transplantation.

This study reviews our clinical experience of patients with polycystic disease. We attempt to predict those patients who will develop renal failure and document benefits of intervention with dialysis and transplantation.

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Patients and methods

Since 1960 78 patients with polycystic kidney disease have been followed up at this hospital. There were 77 adults (48 men and 29 women) with ages from 13 to 74 (mean 39 years) and one boy aged 5. Two sets of non-identical twins were included in the series. Forty-two patients needed dialysis or transplantation. At this centre 14 men, the boy, and nine women received 29 renal allografts. Four men and two women underwent transplantation elsewhere, and a further nine men and three women were maintained on haemodialysis at other centres.

Of the 36 who had not received replacement therapy, 10 are still seen and do not need treatment, five were seen on only one occasion, and nine are dead. Despite careful inquiry 17 were not traced but had been followed for between four months and 12 years (mean five years 5 months) before they were lost to review.

The clinical diagnosis of polycystic kidney disease was suggested by the history and physical examination and confirmed by radiology. Intravenous excretory urography was the method most used, but retrograde urography, renal arteriography, and renal venography were all employed. Combined intravenous urography with nephrotomography was most helpful and was the technique of choice.[6 7] Operative renal biopsy was performed in four patients.

The statistical methods were those of linear regression analysis.

Results

PATHOLOGY

The boy underwent unilateral nephrectomy before transplantation and his kidney had the histological appearances of infantile polycystic disease (Potter type I).[8] Adult polycystic disease (Potter type III) was confirmed by histology in all patients who underwent nephrectomy (nine) or renal biopsy (four).

CLINICAL FEATURES

A definite family history of polycystic kidneys was given by 30 patients. In another four this was less definite. Haematuria, loin pain,

and symptoms attributable to hypertension were the most common modes of presentation (table I). Thirty patients had gross haematuria during their disease, and 18 suffered symptoms of urinary infection with considerable bacterial growth. The average blood pressure at presentation was 160/105 mm Hg (range 120/70-280/170 mm Hg). Of the 63 who were hypertensive (blood pressure over 150/100 mm Hg) 39 were treated with antihypertensive drugs. Four patients had accelerated hypertension (Keith Wagener Barker classification group 3)[9] and one had papilloedema (group 4). Electrocardiography showed left ventricular hypertrophy in 19.

TABLE I—*Presenting symptoms of 78 patients with polycystic disease of the kidneys*

	No (%) of patients
Urinary tract symptoms	51 (65%)
Haematuria	21 (27%)
Loin pain	18 (23%)
Urinary infection	10 (13%)
Azotaemia	8 (10%)
Backache	4 (5%)
Loin mass	3 (4%)
Cardiovascular symptoms	22 (28%)
Hypertension	19 (24%)
Hypertension during pregnancy	3 (4%)
Other presentations	5 (6%)
Coincidental at surgery	3 (4%)
Potential kidney donors	2 (3%)

Some patients had multiple symptoms.

PREGNANCY

There was no information on whether 17 women had had children, though 11 women had. Seven had had their families before polycystic disease was diagnosed, and their pregnancies had been uneventful. One woman with diagnosed polycystic kidney disease had had five pregnancies resulting in one abortion, two neonatal deaths, and two live infants. A further three women developed hypertension, proteinuria, and urinary infection during pregnancy, and investigation showed polycystic kidney disease. The hypertension and proteinuria resolved after pregnancy. Fetal loss was high in these three, but with careful supervision of subsequent pregnancies all had healthy infants. Pregnancy did not appear to accelerate deterioration of renal function or precipitate renal failure.

DETERIORATION OF RENAL FUNCTION IN VARIOUS SUBGROUPS

The development of renal failure was not hastened by pregnancy and urinary infection. Rovsing's operation was performed on 13 patients. End-stage renal failure developed in three patients soon after the procedure, and deterioration of renal function was accelerated in six others.

From our data we believe that it is possible to predict when renal failure will ensue. Patients who developed symptoms early in life developed renal failure needing dialysis at a younger age than those who presented later. These younger patients, however, had a longer interval before entering renal failure than those presenting later. Age at transplantation and age at death were similarly correlated. These trends are highly significant ($P = < 0{\cdot}001$) (fig 1).

TRANSPLANTATION AND HAEMODIALYSIS

Six patients underwent transplantation at other centres. Two died during the first week after operation. Another had two transplants that failed to function and subsequently received haemodialysis for two years. Three other patients are alive with transplanted kidneys that have functioned well for between six and seven years.

At our centre 24 patients underwent renal transplantation and received 29 renal allografts. Twenty-five were from cadavers and four from living related donors, which reflected the hereditary factor in polycystic renal disease. All four patients who received a live donor kidney are alive and well with kidneys that have functioned between one year ten months and nine years eight months (mean four years two months). The 20 patients who received cadaver grafts showed improved graft function at all time intervals compared with 70 patients with other forms of end-stage renal failure who received a cadaver

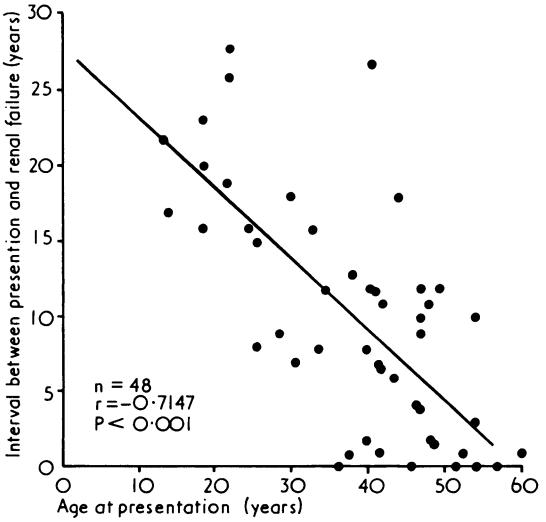

FIG 1—Regression line of interval between presentation and renal failure on age at presentation.

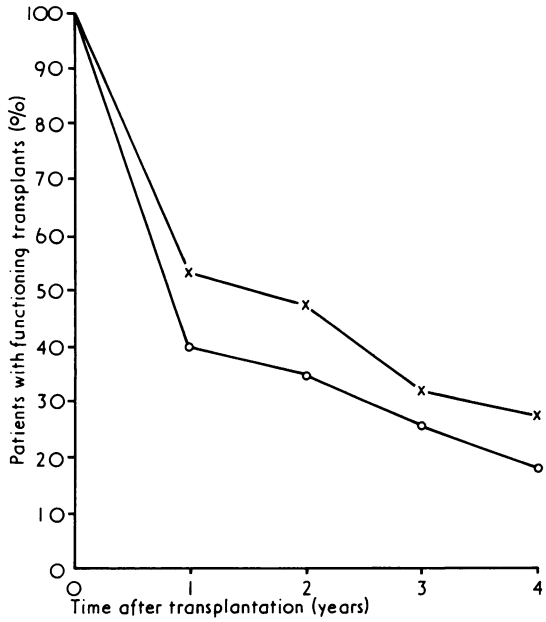

FIG 2—Kidney function after cadaver transplant. Patients with polycystic kidney disease (X) compared to those with renal failure due to other causes (O).

graft during this period (fig 2). Six of these patients remain alive with kidneys that have functioned well for between one year seven months and nine years nine months (mean five years two months). Of the 14 patients who died, nine had a functioning kidney. The causes of death are shown in table II and survival periods in fig 3.

Necropsy showed nine of the 14 had polycystic livers, and four had "berry" aneurysms. Three died from cerebrovascular accidents due to rupture of their aneurysms, but none had problems due to polycystic liver disease.

Twelve patients were maintained on long-term haemodialysis. One died within six months of starting dialysis, and another dialysed in India and has been lost to follow-up. Ten patients remain alive and well between two months and six years of starting dialysis (mean three years five months). Five of 24 patients who underwent transplantation in our unit had unilateral nephrectomy to facilitate transplantation, and in four this was before grafting, but in one it was at the time of transplantation. Two other patients had unilateral nephrectomy before transplantation; one for renal calculi and the other for persistent infection of a cyst.

After transplantation one patient underwent bilateral nephrectomy

TABLE II—*Causes of death after transplantation for end-stage polycystic kidney disease*

Cause of death	Interval between transplantation and death	Renal function
Pneumonia	5 days	Poor
Cytomegalovirus pneumonia	5 days	Poor
Gastrointestinal haemorrhage	2 weeks	Normal
Perforated oesophagus	3 weeks	Normal
Pneumonia and septicaemia	6 weeks	Normal
Perforated colon	2 months	Normal
Gastrointestinal haemorrhage and cerebrovascular accident	2 months 2 weeks	Normal
Gastrointestinal haemorrhage	3 months	Normal
Cerebrovascular accident	10 months	Normal
Myocardial infarction	1 yr 1 month	Nil
Pulmonary candidiasis	1 yr 6 months	Poor
Pneumonia and subacute bacterial endocarditis	2 yr 6 months	Normal
Cerebrovascular accident	2 yr 11 months	Nil
Myocardial infarction	4 yr 4 months	Normal

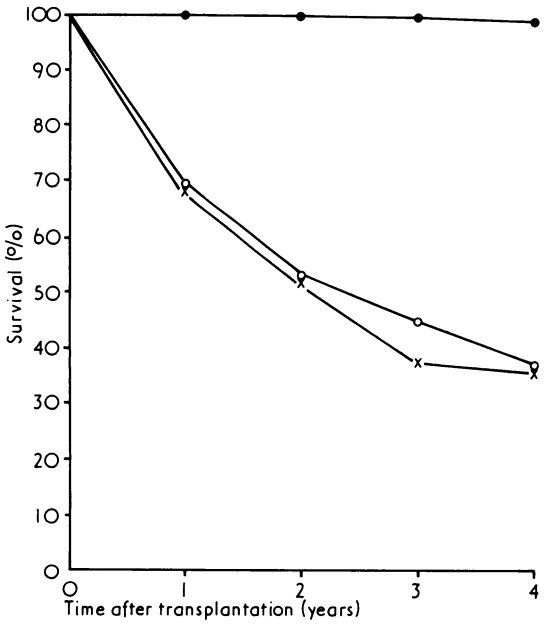

FIG 3—Raw survival after renal transplantation. Three groups of patients are shown, those with polycystic disease who had live donor grafts (●), those with polycystic disease who had cadaver grafts (X), and those who had cadaver transplants for renal failure due to other diseases (O).

for recurrent urinary infection. Haematuria was not significant in those with kidneys still in situ and was not made worse by haemodialysis.

The mean haemoglobin concentration was higher in patients with polycystic kidneys than in a matched group of patients with renal failure from other causes.[10] After successful transplantation, three patients developed erythraemia which did not respond to repeated venesection or treatment with radiolabelled phosphorus. After removal of both polycystic kidneys haemoglobin levels became normal.

HLA TISSUE ANTIGENS

HLA tissue antigens were determined in patients during assessment for transplantation. Certain antigens were more common in patients with end-stage renal failure due to polycystic disease than in the general population. HLA-B7 was present in 46% of these patients but in only 26·1% of the general population, and HLA-A3 was present in 46% of the polycystic patients and 22·5% of the general population.

Discussion

The incidence of adult polycystic renal disease at necropsy is about 1 in 800.[11] Only 25% of these have symptoms during life,[3] but once the disease becomes manifest the renal cysts continue to enlarge, and our data show that end-stage renal failure occurs in over 70% of those who live to the age of 65. This contrasts with infantile polycystic disease, which invariably leads to renal failure in the first few years of life.

In adult-type disease we describe for the first time an important relation between the age at the onset of symptoms and the age of development of terminal renal failure. A young patient with symptoms will develop renal failure early in life, but there is a longer interval before renal failure supervenes than in those patients who present later. For example, a patient who develops symptoms at 20 will develop renal failure near 40. A patient whose disease presents at 50 will develop renal failure near 55. The patient who does not have overt polycystic disease and whose condition is discovered coincidentally or during investigation of the family of a patient with known polycystic disease has a good prognosis. Follow-up may be planned, and for a younger patient annual review should suffice during the early stages of the disease. Older patients may have more rapid development of renal failure necessitating more frequent review. At examination, the blood pressure must be measured, the fundi examined, and blood taken for estimation of electrolyte, urea, and creatinine concentrations. Glomerular filtration rate should be determined, and a chest radiograph and an electrocardiogram obtained. Once the diagnosis has been established, little information can be gained by repeated urography.

It has been suggested that pregnancy is not advisable in women with polycystic kidney disease because of the risk of transmitting the condition to the children and the risk of accelerating deterioration of renal function.[3] Certainly, genetic counselling should be given as the disease is transmitted as an almost pure dominant, and the incidence in progeny nears one in two. We find no evidence to support the view that pregnancy causes more rapid deterioration of renal function. Provided careful supervision is maintained, pregnancy should be uneventful and lead to the birth of a viable infant.

We agree with Lazarus *et al*[12] that Rovsing's operation causes more rapid development of renal failure and think that there is no longer a place for this operation in the modern management of polycystic kidney disease. We confirm that renal transplantation and long-term haemodialysis are effective methods of treating end-stage renal failure in polycystic kidney disease.[10–14] Because polycystic renal disease is hereditary, cadaver donors were used for most patients. Nevertheless, an effort should be made to find a suitable related donor since live donor transplants are much better.[15]

Others have recommended bilateral nephrectomy before transplantation in patients with a history of renal infection or severe haematuria[11 13 14] None of our patients had such preparation, and we had no appreciable problems except for one patient, who needed bilateral nephrectomy for recurrent urinary infections. Indeed, it appears safe and desirable to leave the kidneys in place. The advantages of leaving the kidneys accrue primarily while patients are on haemodialysis for a moderate urinary output is often maintained, which may alleviate the need for severe fluid restriction. A relative erythraemia occurs, and vitamin D metabolism may be more efficient in patients with kidneys since calcium absorption decreases in patients after nephrectomy.[16] The main indications for pretransplant nephrectomy are persistent or recurrent urinary infections and renal calculi associated with these infections, but we found these complications uncommon.

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The nurse's role in immediate postoperative care

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Summary

From the time that a patient leaves the care of the anaesthetist after an operation until he wakes in the ward his physiological state should be continuously and expertly supervised. Postoperative nurses are provided only when the operating theatre has a recovery room. A survey among consultants and nurses in one region showed that many surgical units did not have recovery rooms and that inexperienced ward nurses were often sent to collect patients. The survey showed that most nurses were competent to care for unconscious patients so long as an emergency did not arise. In many hospitals the facilities for the safe nursing of postoperative patients were totally inadequate.

The very least that is needed is good communications with the anaesthetist, adequate lighting, and a source of oxygen and suction. Because of the shortage of nurses it would be realistic to recognise that trainee nurses are likely to have to care for postanaesthetic patients early on and to train them accordingly. Nevertheless, recovery nurses, whose sole responsibility is to care for a patient until he has recovered from anaesthesia, should be appointed for all busy surgical units.

Introduction

In parts of Europe and the United States of America specially trained nurses take an active though limited part in the conduct of anaesthetics. In the United Kingdom they play no part in the specialty. British anaesthetists defend this anomalous position by maintaining that nurses are not qualified to accept the responsibilities that the specialty demands. Yet immediately the patient leaves the operating theatre his care passes into the hands of a nurse, who may be a fully trained sister or a first-year trainee. Although the anaesthetist remains responsible for the patient, he is invariably not readily available to deal with any emergency that may arise in the patient who has just passed out of his hands.

It is a tribute to modern anaesthesia and postoperative nursing care that only about 100 unexplained deaths associated with anaesthesia occurred yearly in 1959-70,[1] while over 4 000 000 anaesthetics were given each year in England and Wales alone. The Registrar General's figures do not identify the stage when death occurred, nor do they distinguish between avoidable and unavoidable deaths. Wylie recently examined the Medical Defence Union records of the past 20 years and reported that about a third of the complications associated with anaesthesia occurred in the postoperative period, about half of these culminating in cardiac arrest.[2] These figures suggest that failure in postoperative management may have been responsible.

Immediately after operation patients are exposed to certain risks such as regurgitation and vomiting, obstruction of the respiratory passages, and cardiac insufficiency. Well-trained staff and adequate resuscitation equipment are essential for treating, and whenever possible preventing, these complications. The equipment is best provided in a modern recovery room within easy reach of the anaesthetist.[3] Theatre nurses are officially provided to care for postanaesthetic patients only if there is a recovery room attached to the theatre. If there is no such room the theatre nurse usually relinquishes her responsibility for the patient to a ward nurse as soon as possible. The ward nurse may be inexperienced, frightened, and uncertain of her role in these circumstances. And, because there is no recovery room, she may have to care for the patient in a poorly lit, draughty corridor while the anaesthetist is by this time busy with the next case. The continuing shortage of nurses, the ever-increasing volume of surgery,[4] and the lack of special equipment are making it increasingly difficult to provide adequate care in the critical postanaesthetic period.

Most anaesthetists are aware of these problems and modify their techniques so that patients will wake promptly at the end of surgery and rapidly regain their reflexes. This is not always in the best interests of the patient, who might benefit from a peaceful recovery in a safe environment. The patients who take the longest time to wake are those who have had an inhalation anaesthetic (as distinct from a muscle relaxant artificial ventilation technique) lasting half an hour or more. These anaesthetics are given for such common and relatively simple procedures as herniorrhaphy and varicose vein surgery in otherwise fit people, and it is often these patients who unexpectedly provide the most problems in the recovery period.

Although they are trained in their part in the management of cardiorespiratory arrest, nurses are not generally sufficiently aware of the particular complications that arise in the immediate postoperative period and how they can be prevented. Records of complications, even of cardiac arrest, are often inadequate, and in many cases nothing is recorded in the patient's notes if resuscitation has been successful.

We decided to investigate this further, concentrating on nursing knowledge and competence and on the facilities provided for postoperative recovery in a cross-section of hospitals in this region.

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