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Angiographic diagnosis of polycystic renal disease

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POLYCYSTIC disease of the kidney usually presents no difficulty in diagnosis. The presence of arterial hypertension, palpable kidneys, family history of renal disease, various degrees of renal insufficiency, and urographic and pyelographic evidence of multiple, bilateral renal masses establish the diagnosis in the majority of patients. Problems in diagnosis arise when the combination of findings is absent.[1] In this problem group are those patients with arterial hypertension, or renal insufficiency, or both; those relatively symptom-free but with minor abnormalities disclosed by intravenous urography or retrograde pyelography; and those thought to have renal tumors or solitary cysts from evidence on pyelograms.

The contribution of selective renal angiography to the diagnosis of polycystic renal disease is the subject of this report. For the purpose of this series, polycystic disease is defined as a familial and probably congenital pathologic process in which numerous cysts are located diffusely throughout the renal parenchyma of both kidneys, often associated with arterial hypertension, various degrees of renal insufficiency, and occasionally cystic disease of the liver. An elaborate distinction between so-called multicystic disease and polycystic renal disease is not included in this report.

MATERIALS AND METHODS

From a series of 1,500 selective renal arteriograms, a diagnosis of polycystic renal disease was established in 14 patients. These patients each had been studied because of a suspected renal mass (not polycystic disease), for the evaluation of arterial hypertension or of uremia or of both disorders. In some instances urographic abnormalities had prompted the angiographic study.

All patients were studied by means of intravenous pyelography, and selective renal angiography.[2] A premolded, medium-sized, green polyethylene Ödman-Ledin catheter† was introduced through a percutaneous femoral puncture, by means of the Seldinger[3] technic. Depending on the size

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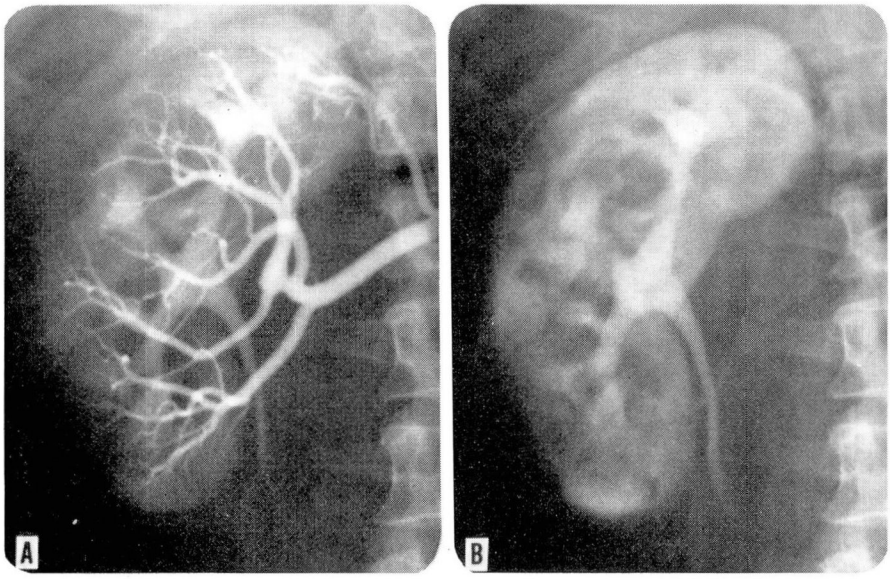

Fig. 1. A, Arterial phase of the selective injection of the right renal artery, showing displacement of the interlobar arteries and the mid portion of the right kidney. B, Nephrographic phase in the same patient, showing numerous nonopacified patches produced by multiple cysts, many of which do not produce arterial displacement.

of the renal arteries, from 4 to 8 ml of sodium diatriozate* was selectively introduced into each renal artery. Aortograms of each patient were made after injection of from 20 to 30 ml of the contrast medium.

RESULTS

Of the 14 patients, seven were men ranging in age from 20 to 74 years; the age range of the seven women was 17 to 60 years. Nine patients were hypertensive. Blood urea levels and creatinine clearance studies of six patients showed compromised renal function. Only three patients had family histories of polycystic renal disease. The kidneys were palpable in only two of the patients.

Radiorenography was performed in four patients, and abnormalities were disclosed in three. The angiographic diagnosis of polycystic renal disease in those patients was based upon abnormalities either in the arterial phase or in the nephrographic phase of the renal angiograms. Bilateral disease in all the patients was demonstrated.

Displacement of interlobar and segmental arteries (*Fig. 1A*) was found in 12 patients in whom large cystic masses were present. The nephrographic

* *Hypaque sodium 50%, Winthrop Laboratories.*

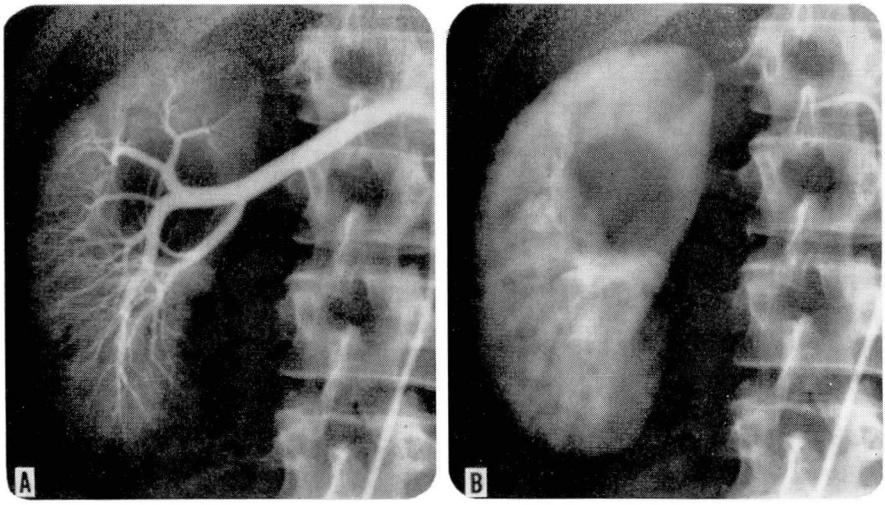

Fig. 2. A, Arterial phase of the selective injection of the right renal artery, showing minimal displacement of the segmental branch of the posterior interlobar artery in a patient thought to have a solitary renal mass. B, Nephrographic phase in the same patient, showing numerous defects throughout the kidney, ranging in size from a few millimeters to several centimeters.

phase of the angiograms of these patients showed no opacification in the regions of the cysts (*Fig. 1B*).

In the two patients in whom small cysts were demonstrated, displacement of major arteries was not a prominent finding (*Fig. 2A*). In these patients, the nephrograms provided the key to the diagnosis. Defects from a few millimeters in diameter to from 1 to 2 cm were distributed throughout both kidneys, producing a spongelike or "Swiss cheese" appearance. It was the urograms and pyelograms of these patients which did not demonstrate polycystic renal disease (*Fig. 3*).

Moderate to severe enlargement of kidneys was often masked by poor demonstration of the pelviocaliceal system because of decreased renal function or predominant cystic change in the collecting system. Both the arterial and the nephrographic phases of the arteriogram provided important evidence of renal enlargement.

The volume of renal parenchyma could be estimated from the relative size of renal outline as compared with the size of the renal arterial tree. A small arterial system supplying a large kidney indicated a significant loss in renal parenchyma (*Fig. 4*).

Associated abnormalities in the main renal arteries were found in three patients. In one patient, there was a relatively nonocclusive atherosclerotic

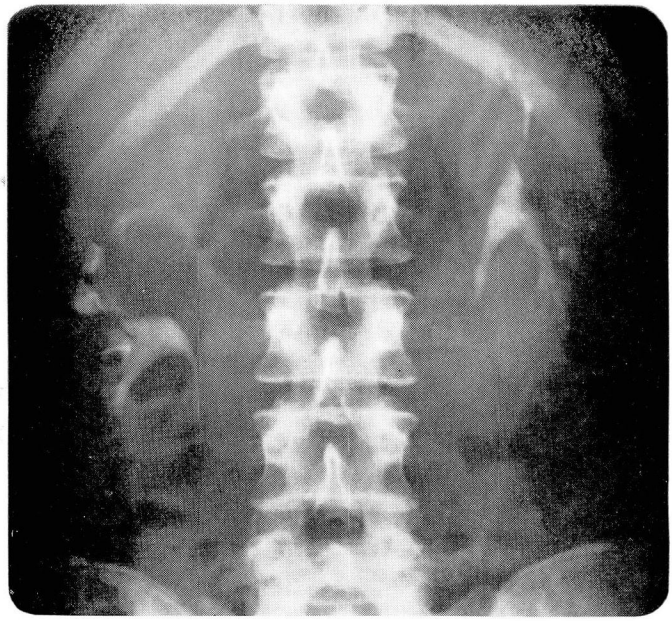

Fig. 3. Five-minute film of the intravenous urogram of the patient shown in *Figure 2*, showing a large kidney but only a localized pressure defect on the upper calyxes of the right kidney in the region of the large cyst. There was no displacement of the pelvis or calyxes on the left side.

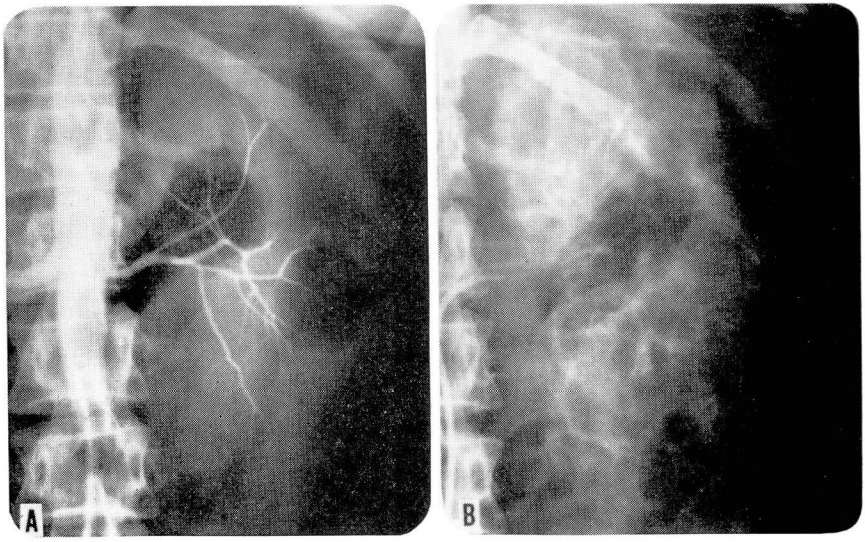

Fig. 4. A, Arterial phase of the selective injection of the left renal artery, showing a small arterial supply to the left kidney, despite the large size of the kidney as shown in the nephrographic phase. B, Nephrographic phase of the angiogram showing that the majority of the renal parenchyma has been replaced by cysts.

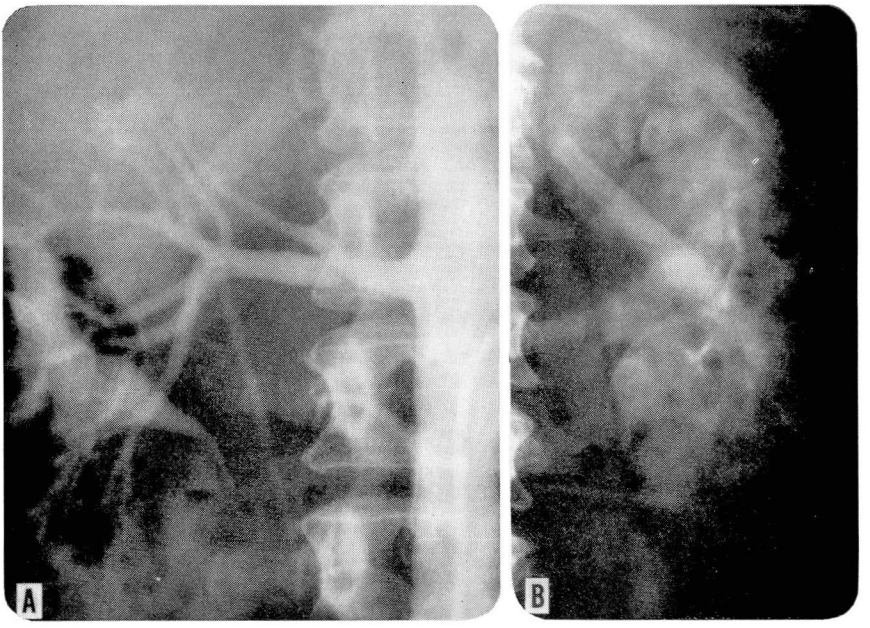

Fig. 5. A, Arterial phase of the angiogram, showing an atherosclerotic plaque at the bifurcation of the right renal artery. B, Nephrographic phase of the selective injection of the left renal artery, showing irregular outlines due to numerous small cysts throughout the kidney.

lesion; in one patient, moderately extensive medial fibroplasia[4] affecting an upper pole branch of the right renal artery; and in one patient, a moderately stenosing atherosclerotic lesion of the main right renal artery (*Fig. 5*).

Intravenous pyelograms showed no abnormality in each of three patients, evidence of unilateral renal mass in each of five, bilateral mass in each of three, and no demonstration of kidneys in three. Retrograde pyelograms of two patients revealed no abnormality.

DISCUSSION

In consideration of the results it is important to emphasize that before selective angiography, of the 14 patients only three showed definitive evidence of polycystic renal disease.

The six patients investigated for arterial hypertension are of special interest, because in four of them polycystic renal disease was an unexpected finding, as was also the coexistence of arterial disease. Stenoses were moderately occlusive in two patients. One patient had extensive involvement by medial fibroplasia,[4] and the other an atherosclerotic plaque. In the

latter patient, the blood pressure returned to normal levels after endarterectomy.

The usefulness of selective renal angiography in the preoperative evaluation of possible renal tumors or cysts was demonstrated in five patients who had urographic or pyelographic evidence of solitary lesions. The demonstration of bilateral multiple cysts by these roentgenographic technics made surgical exploration unnecessary.

The diagnosis of polycystic disease is difficult when the cysts are small. This difficulty is emphasized by our finding that the only evidence for polycystic renal disease in nine (of 14) patients was enlarged kidneys. The importance of abnormally large kidneys in the diagnosis of polycystic renal disease has been emphasized by Lalli,[5] and is apparent in our series of patients, each of whom had a kidney longer than 13.5 cm.

The size of the renal arterial system corresponded with the volume of renal tissue, as judged by the nephrographic phase of the angiogram. The great disproportion between a large-sized polycystic kidney and a small arterial system provides a useful index of volume of renal parenchymal tissue. Our series of patients is too small to determine whether or not the discrepancy has prognostic value.

SUMMARY

The arteriographic findings in 14 patients with polycystic renal disease are presented. This diagnosis was not suspected in nine of the patients before they underwent angiography. Abnormalities in the nephrographic phase of the angiograms offered the most consistent evidence. No deformity of the pelviocaliceal system in each of three patients was demonstrated on intravenous urograms or retrograde pyelograms. Three hypertensive patients were found to have coexistent renal arterial disease, and in one of these patients relief of hypertension occurred after endarterectomy of an atherosclerotic plaque of the main renal artery.

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