Value of antenatal diagnosis of abnormalities of the urinary tract

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Abstract

Objective—To assess the value of antenatal diagnosis of abnormalities of the urinary tract on ultrasonography.

Design—Retrospective study.

Setting—Two obstetric units in Glasgow.

Subjects—62 Fetuses in which renal abnormalities were diagnosed on antenatal ultrasonography.

Interventions—Six fetuses had their bladders aspirated to determine renal function. Fifteen pregnancies were terminated on the basis of the findings on antenatal ultrasonography, and if possible necropsy was performed on the fetuses. In babies who were born alive the final diagnosis was made by postnatal ultrasonography, intravenous urography, radionuclide scanning, cystography, and, in those who died in the early neonatal period, necropsy. Neonates who were referred with a known obstructed kidney had nephrostomy or pyeloplasty.

End point—Assessment of the value of antenatal diagnosis of renal abnormalities on ultrasonography for babies who had no clinical evidence of disease postnatally.

Main results—Eighteen fetuses did not survive birth; the antenatal diagnosis was accurate in all 18. Of the 44 babies born alive, five had normal urinary tracts, in two of whom antenatal ultrasonography had probably indicated a false positive diagnosis. Fourteen babies died during the early neonatal period. Twenty five babies with renal abnormalities were followed up; the antenatal diagnosis was inaccurate for 10 of them, the commonest misdiagnosis being hydronephrosis for multicystic kidney and vice versa, and there was one false positive diagnosis. The initial clinical findings in 14 babies would have led to the early detection of a urological abnormality. In the 30 babies with no clinical evidence of disease the antenatal diagnosis was of definite value in eight, probable value in 15, and marginal value in seven. Overall, an accurate antenatal diagnosis was made in 46 of the 62 cases (74%); in 12 cases renal disease was detected but its specific nature was not determined; and in four cases the diagnosis was misleading.

Conclusions—The overall value of antenatal diagnosis is that it indicates early termination of fetuses with fatal renal disease, prepares parents and medical staff for the likelihood of serious neonatal problems, and shows abnormalities of the urinary tract that may not be detected postnatally.

Introduction

The incidence of abnormalities of the urinary tract that can be diagnosed antenatally has been estimated as 1/1000 live births.¹ Such diagnoses raise new and possibly controversial questions and have implications for the fetus or neonate, or both, and for the parents

and the nursing and medical team. In a study of disorders that were diagnosed antenatally by ultrasonography and required an operation we found that urological abnormalities comprised 34%.² We now report cases of such abnormalities that we have encountered during the past four and a half years.

Methods

We detected 46 cases of fetal uropathy on ultrasonography in two large obstetric units in Glasgow during 54 months (August 1983 to February 1988), and 16 pregnant women were referred from other district maternity units in west Scotland after fetal uropathy was diagnosed on ultrasonography. Antenatal bladder aspirations guided by ultrasonography were performed on six fetuses to determine renal function. One fetus had a renal cyst aspirated, but no fetus received intrauterine drainage. In those pregnancies that were terminated necropsy of the fetus was performed if possible.

In babies who were born alive the final diagnosis was made by postnatal ultrasonography, intravenous urography, radionuclide scanning, cystography, and, in those who died in the early neonatal period, necropsy. Neonates who were referred with a known obstructed kidney had nephrostomy or pyeloplasty during the neonatal period. In other cases an operation was deferred after assessment or was not considered necessary. The value of antenatal diagnosis of renal abnormalities for their subsequent management was classified as marginal (for example, for mild unilateral dilatation and agenesis of one kidney); probable (for example, for unilateral obstruction of the pelviureteric junction and multicystic kidney); or definite (for example, for treatable conditions affecting both kidneys, such as posterior urethral valves).

Results

Eighteen fetuses (29%) did not survive birth (table). Fifteen of the pregnancies were terminated on the basis of findings on antenatal ultrasonography, which had been done at a mean gestational age of 18 (range 14-23) weeks and agreed with the findings at necropsy in all cases. Eight fetuses had associated congenital abnormalities, including five with prune belly syndrome, and antenatal aspirations of the bladder confirmed poor renal function in four. Two fetuses that were not terminated (one died in utero and one was stillborn) had lethal urological abnormalities. One fetus with unilateral cystic dysplasia aborted spontaneously.

Forty four babies were born alive. Five of these had normal urinary tracts; in two of them (cases 19 and 23) ultrasonography had probably given a false positive diagnosis as follow up did not show any renal abnormalities. Three babies (cases 20-22) had had unilateral or bilateral hydronephrosis on repeated antenatal

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Br Med J 1989;298:1417-9

scanning, but this decreased towards term and at birth and consequently their upper urinary tracts were normal; we regarded these babies as having had transient intrauterine hydronephrosis, not as having yielded a false positive diagnosis.

Fourteen babies died during the early neonatal period. The abnormalities in these cases had generally been diagnosed later than those in the terminated pregnancies (mean gestational age 29 (range 17-38) weeks). If these abnormalities had not been diagnosed in utero urological problems would probably have become apparent in most of the babies after delivery. Only one fetus in this group had had a bladder aspiration. This had confirmed poor renal function at 27 weeks. The antenatal diagnoses were inaccurate in four cases, but this did not affect the outcomes. Eight of the babies had associated congenital abnormalities.

Twenty five babies with renal abnormalities were followed up. The antenatal diagnosis had been made at

a mean gestational age of 29 (range 18-39) weeks, 18 having been diagnosed in the third trimester. The antenatal diagnosis was accurate in 15 cases but inaccurate in 10; the commonest misdiagnosis was hydronephrosis mistaken for multicystic kidney or vice versa. There was one false positive finding (case 62), when a normal left kidney (the right kidney was absent) was misdiagnosed as a multicystic right kidney.

The initial clinical findings in 14 of the 44 babies who were born alive would have led to early detection of a urological abnormality. In the 30 others, however, there was no clinical evidence of renal disease. The 30 babies were subdivided to assess the value of antenatal ultrasonography in directing the management of the renal abnormality after birth. The antenatal diagnosis was of definite value in eight of them, of probable value in 15, and of marginal value in seven (including the five babies who had normal urinary tracts after birth).

Overall, an accurate antenatal diagnosis was made in 46 of the 62 cases (74%). In 12 cases renal disease was detected but its specific nature was not determined; and in four cases the diagnosis was misleading, but in no case did this affect the clinical outcome.

Discussion

The overall diagnostic accuracy of antenatal ultrasonography of 74% is similar to that found in comparable series.^{1,3} The main difficulty was distinguishing between dysplastic, hydronephrotic, and cystic kidneys, and this accounted for most of the inaccurate diagnoses. The diagnosis of renal agenesis is also difficult as the possibility of ectopic renal tissue has to be excluded.

No false positive diagnoses of fetal uropathy were made in any of the pregnancies that were terminated in this series, and all but two fetuses had a fatal abnormality of the urinary tract. Aspiration of the fetal bladder to confirm poor renal function was valuable and this may facilitate the decision as to whether to offer termination when the fetal abnormality is considered incompatible with life. Among the babies born alive there were three false positive diagnoses. This did not lead to adverse management in any case, though this is obviously a risk.

Fourteen (32%) babies died during the early neonatal period. In two of these, who had posterior urethral valves, renal function might have been improved by intrauterine drainage, but there is evidence that survival is no better after such procedures, which have an unacceptably high rate of complications.^{4,5} We believe that an early operation after birth to improve the drainage of the urinary tract, particularly for obstruction of the pelviureteric junction, results in reduced parenchymal loss and preserves renal function.⁶ The mortality and the subsequent renal function in the babies in our series are comparable with those in other series with and without fetal intervention.^{1,7,8}

The overall value of antenatal diagnosis in this study was that it indicated early termination of fetuses with fatal disease (15 cases), prepared parents and attendant nursing and medical staff for the likelihood of serious neonatal problems (there were 14 deaths in the early neonatal period), and made staff aware of abnormalities of the urinary tract in neonates without obvious clinical evidence of an abnormality postnatally (30 cases). A minority of fetuses (14) progressed to normal birth and survived with an obvious abnormality that would inevitably have been diagnosed clinically and whose management was not influenced by antenatal diagnosis. Even so, the parents' readiness for postnatal events is important psychologically and socially.

We conclude that once an abnormality of the fetal urinary tract is detected pregnant women should

MINIPRINT TABLE

Diagnosis on antenatal ultrasonography and final diagnosis in fetuses with renal abnormalities that did not survive birth (cases 1-18); in babies born with a normal renal tract (cases 19-23); in babies with renal abnormalities who died early in neonatal period (cases 24-37); and in babies with renal abnormalities who survived neonatal period. (Pregnancy was terminated in cases 1-15)

Case No	Sex	Age at antenatal diagnosis (weeks)	Antenatal diagnosis	Final diagnosis
FETUSES THAT DID NOT SURVIVE BIRTH				
1	M	14	Distended bladder, hydroureters	Urethral atresia
2	M	15	Abdominal cysts	Urethral atresia, prune belly syndrome
3	M	19	Polycystic kidneys	Polycystic kidneys
4	M	16	Distended bladder, bilateral hydronephrosis, dysplastic kidneys	Urethral valves, dysplastic kidneys
5	F	21	Polycystic kidneys	Bilateral renal cystic dysplasia
6	M	17	Distended bladder, hydroureters, hydrocephalus	Urethral, oesophageal, duodenal, and anorectal atresia, hydrocephalus
7	M	21	Distended bladder, bilateral hydronephrosis	Megalocystis, megaloureter, prune belly syndrome
8	F	16	Prune belly syndrome	Urethral atresia, bilateral renal dysplasia
9	F	17	Prune belly syndrome*	Prune belly syndrome
10	M	21	Small kidneys	Bilateral renal agenesis
11	M	17	Bilateral renal agenesis	Bilateral renal agenesis
12	M	18	Prune belly syndrome†	Prune belly syndrome
13	M	21	Abnormal renal mass, hydrocephalus	Horseshoe kidney megalocystis, megaloureter, hydrocephalus, myelomeningocele
14	M	23	Polycystic kidneys	Polycystic kidneys (Meckel's syndrome)
15	M	18	Distended bladder, bilateral hydronephrosis*	Urethral valves
16†	M	14 (23‡)	Prune belly syndrome	Prune belly syndrome
17§	M	20 (24‡)	Multicystic left kidney	Cystic dysplasia of right kidney
18¶	M	33 (34‡)	Bilateral renal agenesis	Bilateral renal agenesis
BABIES BORN WITH A NORMAL RENAL TRACT				
19	F	40	Hydronephrosis of left kidney, left hydroureter	Normal
20	F	22	Bilateral hydronephrosis	Normal
21	M	33	Hydronephrosis of right kidney	Normal
22	M	23	Hydronephrosis of right kidney	Normal
23	F	24	Cystic kidneys	Normal
ANTENATAL DIAGNOSIS ACCURATE				
24	M	27	Distended bladder, bilateral renal dysplasia	Urethral atresia, bilateral renal dysplasia
25	M	20	Abnormal renal outline	Polycystic kidneys
26	M	34	Bilateral renal agenesis	Bilateral renal agenesis, oesophageal atresia, pulmonary hypoplasia
27	F	35	Bilateral renal agenesis	Bilateral renal agenesis, myelomeningocele
28	F	25	Cystic kidneys, bilateral hydroureters	Cystic dysplastic kidneys, bilateral hydroureters, urethral valves
29	M	32	Small kidneys	Bilateral renal agenesis (Potter's syndrome)
30	M	25	Polycystic kidneys	Polycystic kidneys (Meckel's syndrome)
31	F	38	Hydronephrosis of left kidney	Mild hydronephrosis of left kidney (Möbius's syndrome)
32	F	32	Distended bladder, bilateral hydronephrosis	Urethral valves
33	F	24	Prune belly syndrome	Prune belly syndrome
ANTENATAL DIAGNOSIS INACCURATE				
34	M	27	Distended bladder, bilateral renal agenesis	Urethral valves
35	F	33	Polycystic kidneys	Renal dysplasia
36	F	32	Hydronephrosis of right kidney, hydrocephalus	Right ureterocele, bilateral renal dysplasia, hydrocephalus, duodenal atresia, imperforate anus
37	F	17	Bilateral renal agenesis	Pelvic horseshoe kidney, laryngeal atresia
ANTENATAL DIAGNOSIS ACCURATE				
38	M	32	Hydronephrosis of right kidney, right hydroureter	Hydronephrotic left upper moiety of duplex system, left ureterocele
39	M	24	Bilateral hydronephrosis	Prune belly syndrome, talipes equinovarus
40	M	27	Bilateral hydronephrosis	Bilateral obstruction of pelviureteric junction
41	M	28	Hydronephrosis of right kidney	Obstruction of right pelviureteric junction
42	M	38	Bilateral hydronephrosis	Penile urethral bands
43	M	31	Enlarged right kidney	Right renal hypertrophy, left renal agenesis
44	F	34	Hydronephrotic left upper moiety of duplex system	Hydronephrotic left upper moiety of duplex system
45	F	18	Distended bladder, bilateral hydronephrosis	Megalocystis, megaloureter, obstruction of right pelviureteric junction
46	M	29	Multicystic right kidney	Multicystic right kidney
47	F	35	Hydronephrosis of left kidney	Obstruction of left pelviureteric junction
48	M	32	Hydronephrosis of left kidney	Obstruction of left pelviureteric junction
49	M	38	Hydronephrosis of left kidney	Obstruction of left pelviureteric junction
50	M	36	Hydronephrosis of right kidney	Right ureterocele
51	M	38	Distended bladder, bilateral hydronephrosis	Megalocystis, megaloureter, duplex left ureter
52	F	21	Hydronephrotic right upper moiety and ureterocele	Hydronephrotic right upper moiety and ureterocele
ANTENATAL DIAGNOSIS INACCURATE				
53	F	18	Multicystic right kidney	Dysplastic right kidney
54	M	24	Bilateral hydronephrosis, bilateral hydroureter	Hydronephrosis of left kidney, left hydroureter, multicystic right kidney
55	M	17	Bilateral hydronephrosis*	Bilateral renal dysplasia
56	M	36	Bilateral hydronephrosis	Left ureterocele, hydronephrosis of left kidney, dysplastic right kidney
57	M	19	Hydronephrosis of left kidney	Multicystic left kidney
58	M	39	Multicystic left kidney	Obstruction of left vesicoureteric junction
59	F	30	Multicystic right kidney	Obstruction of right pelviureteric junction
60	F	37	Bilateral renal cysts	Bilateral obstruction of pelviureteric junction
61	F	18	Bilateral renal cysts	Obstruction of right vesicoureteric junction
62	F	37	Multicystic right kidney	Right renal agenesis, normal left kidney (Klippel-Feil syndrome)

*Bladder aspiration performed.

†Intrauterine death.

‡Figures in parenthesis denote age at which fetus was delivered.

§Spontaneous abortion.

¶Stillbirth.

be referred to a regional centre for confirmation of the diagnosis and be monitored subsequently. The parents and medical team should discuss the management of the abnormality. Antenatal diagnosis inevitably raises moral and ethical questions, and only frequent audit of the implications and outcomes of such practices will enable clinicians to formulate the best management policies.

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(Accepted 31 March 1989)

Bilateral fetal uropathy: What is the outlook?

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Abstract

Objective—To assess the morbidity and mortality associated with a prenatal diagnosis of bilateral fetal uropathy.

Design—Retrospective study.

Setting—Departments of radiology, paediatric surgery, obstetrics, and pathology in two teaching hospitals that serve as referral centres for the Yorkshire region.

Patients—126 Cases of fetal uropathy were referred either prenatally or postnatally from hospitals in Yorkshire between August 1982 and December 1987. The disease was bilateral in 54 cases and unilateral in 72 cases. In 14 cases bilateral fetal uropathy was associated with co-existent disease.

Interventions—All cases were managed individually by an obstetrician after discussion with the radiologists and paediatric surgeons. Babies who survived were treated prophylactically with antibiotics after delivery and were operated on if appropriate.

End point—Assessment of prognosis for long term renal function for each baby referred between August 1982 and December 1987: follow up ranged from six months to five years.

Measurements and main results—Of the 54 fetuses with bilateral fetal uropathy, 13 were terminated as the prenatal findings of ultrasonography were considered to be incompatible with long term survival. Ten of the liveborn babies died, five of renal or pulmonary insufficiency, or both, and five of associated congenital anomalies. Thirty one infants survived to follow up; four of these had serious coexistent disease and two had impaired renal function. Thus the overall mortality was 43% and the morbidity rate 19%. The renal anomaly was associated with other serious disease in 14 cases (26%) compared with two (3%) of the 72 cases of unilateral fetal uropathy. All but two of the 27 infants with isolated bilateral urinary tract disease had excellent prospects for survival.

Conclusion—Although bilateral fetal uropathy is associated with a high morbidity rate and mortality, careful prenatal assessment can help to identify fetuses with a poor prognosis. The outlook for a fetus with isolated renal disease if treated promptly after delivery is excellent and compares favourably with that reported after prenatal surgical intervention.

Introduction

Detection of fetal urinary tract anomalies may be valuable for both prenatal and postnatal management,

particularly when the abnormality is bilateral.¹ The potential benefit is, however, offset by the poor prognosis often associated with bilateral fetal uropathy. Doctors had hoped that the introduction of fetal surgery would help to reduce the morbidity and mortality associated with this disease by preventing pulmonary hypoplasia and progressive impairment of renal function. Several recent studies have discussed the possible benefits of decompressing the fetal urinary tract surgically.²⁻⁴ Though the authors generally accepted that intervention is not justified for unilateral disease, they disagreed about the management of bilateral disease. We carried out a study to establish the morbidity and mortality associated with bilateral fetal uropathy, with particular reference to the incidence of coexistent congenital abnormalities and the long term outlook for renal function.

Patients and methods

Between August 1982 and December 1987, 126 cases of fetal uropathy were referred to our units either prenatally or postnatally for further assessment. The disease was bilateral in 54 cases; 28 of these were from our own districts, and 26 were referred from other hospitals throughout the Yorkshire region. Most of the anomalies had been detected by routine antenatal ultrasonography, which is normally performed at 17-19 weeks of gestation. There was no fixed policy for following these cases as generally we considered that each case should be managed individually and that the severity of the abnormality should be taken into consideration.

We reviewed the prenatal and postnatal records, including necropsy reports if appropriate, and assessed the incidence of congenital abnormalities in fetuses with unilateral and bilateral uropathy. Cases of equivocal dilatation of the urinary tract were omitted. Routine ultrasonography is performed on at least 95% of all pregnant women in Leeds, allowing us to estimate the overall incidence of fetal anomalies in the population.

Results

During the study 46 775 pregnancies were registered in Leeds. We saw 78 fetuses with an important urinary tract anomaly; the disease was bilateral in 28 of these. This gave an incidence of all anomalies of the urinary tract of 1 in 600 pregnancies and an incidence of bilateral fetal uropathy of 1 in 1670 pregnancies.

Thirteen of the 54 fetuses with bilateral uropathy were terminated as the antenatal findings were incompatible with long term survival; eight had a primary urinary tract anomaly and in five the uropathy

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Br Med J 1989;298:1419-20