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Research Article

Urinary System anomalies at birth

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ABSTRACT

Background: Congenital anomalies of urinary system are common and are found in 3-4% of population, and lethal urinary anomalies account for 10% of termination of pregnancy.
Methods: A study was done to know the incidence of congenital anomalies at birth for the period of 4 months from May 99 - Sept 99 at Cheluvamba hospital attached to Mysore medical college. Congenital anomalies in the still births, live births and aborted fetuses >20 weeks were studied along with the case history and ultrasound reports. Aborted fetuses and still born babies were collected for autopsy after the consent of parents. These babies were fixed in 10% formalin and autopsy was done after fixing, and anomalies were noted.
Results: Total births during study period were 3000. There were 61 babies with congenital anomalies and 6 babies had anomalies of urinary system. Among the urinary system anomalies 1 baby had bilateral renal agenesis, 1 baby had unilateral renal agenesis with anophthalmia (Fraser syndrome), 2 babies had Multicystic dysplastic kidney disease (MCDK) and 1 live baby had hydronephrosis due to obstruction at pelvi ureteric junction, and 1 live female baby had polycystic kidneys.
Conclusion: Incidence of urinary system anomalies in the present study was 2 per 1000 births. U/S detection of urinary anomalies varies with period of gestation, amniotic fluid volume and visualisation of urinary bladder. Autopsy helps to detect renal agenesis.

Keywords: Hydronephrosis, MCDK, Omphalocele, Anophthalmia, Renal agenesis, Ultrasound (U/S)

INTRODUCTION

About 10% of all new-borns have developmental anomalies of the urinary system. Most of these anomalies do not cause clinical problems. About 45% of childhood renal failure, however result from anomalous development of ureteric bud or metanephros. Since the development of these anlagen is dependent on inductive signal from the other. The kidney may fail to develop on one or both sides. Infants with bilateral renal agenesis are stillborn or die soon after birth. In contrast babies with unilateral renal agenesis often live due to compensatory hypertrophy. For unknown reason 75% of infants with renal agenesis are male.[1]

Relative frequency of unilateral and bilateral renal agenesis is difficult to determine because unilateral agenesis often goes undetected. Autopsy data suggest that unilateral agenesis is 4-8 times more than bilateral. Bilateral renal agenesis is associated with severe oligoamnios and consequent Potters syndrome, includes deformed limbs, wrinkly dry skin & abnormal face, due to uterine wall compression over the growing fetus due to reduced amniotic fluid.[1]

Unilateral renal agenesis is usually associated with different spectrum of anomalies, including complete absence of paramesonephric duct derivatives in females, heart defects and abnormal constrictions of gastro intestinal tract.[1]

Lethal urinary anomalies account for 10% of terminations. Detection of nonlethal anomalies is influenced by many factors, such as timing of the ultrasound examination, experience of the sonologist and quality of the equipment. Normal amniotic fluid volume (AFV) in the 2nd half of the pregnancy suggests at least one functioning kidney and patent urinary conduit to amniotic cavity. Oligoamnios if present, suggest strong suspicion of urinary tract anomalies.[2]

Although prenatal diagnosis of anomalies of urogenital system is relatively easy based on ultrasonographic images, corresponding to the dilated urinary tract, using the U/S finding, to provide prognostic counselling to the couple is a difficult task. Counselling becomes a dilemma as many anomalies may manifest late in the 2nd trimester. Data on antenatal diagnosis and postnatal follow up is important for assessing the prognosis.[3]

The development of urinary system takes place from the intermediate mesoderm along the posterior wall of the abdominal cavity. The growth begins around the 4th post conception week and is complete by 12th week. Routine U/S has become a part of the obstetric care and it is being increasingly used to detect the congenital malformations. Though U/S & MRI are valuable tools, a fetal autopsy after termination of pregnancy is essential to confirm findings and to arrive at definite diagnosis. Fetal autopsy can give significant additional information and it is helpful in identifying the cause of fetal loss.[4]

METHODS

A study was done to know the incidence of congenital malformations in Cheluvamba Hospital attached to Mysore Medical College, from May 1999 - Sept 1999. During the study period of 4 months, there were 3000 births. Study of still born babies, fetuses >20 weeks and live born babies with congenital anomalies was done. There were 61 babies with congenital anomalies, 19 were live births and 42 were abortions and still births. Details of antenatal reports were collected. Twenty six babies were collected for autopsy after the parents' consent. Babies were fixed in 10% formalin solution and autopsy was done after one week.

RESULTS

There were 61 babies with different congenital anomalies and 6 babies had anomalies affecting the renal system. 4 babies were still born and autopsy was done and 2 were live babies and U/S done to confirm findings of antenatal ultrasonography reports.

There was one case of bilateral renal agenesis associated with anophthalmia and low ano rectal anomaly.

Case No. 1: A male baby weighing 2.4 kg was born to 19 year old gravida 2 para 1 living 1 at 32 weeks gestation, and H/o 2nd degree consanguinity. U/S report showed gross oligoamnios with signs of intrauterine death. Autopsy report showed bilateral renal agenesis, anophthalmia & low ano rectal anomaly and features were similar to Fraser syndrome.

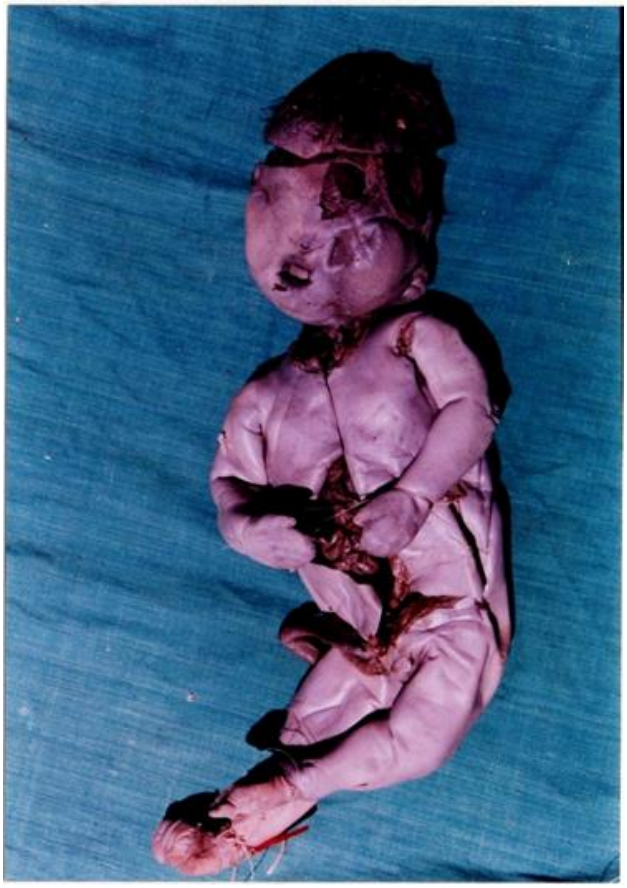

Figure 1: Fraser syndrome: bilateral anophthalmia, bilateral renal agenesis.

Unilateral renal agenesis is 3-4 times more common than bilateral agenesis, and is seen in 1in 1000 births. U/S shows normal amniotic fluid volume and visualisation of urinary bladder. In the present study there was one case of unilateral renal agenesis with other system involvement.

Case No. 2: A still born female baby weighing 1.75 kg was born to primi gravida aged 21 years, at 32 weeks gestation and there was no h/o consanguinity. U/S showed dilated lateral ventricles, large sacral meningocele, omphalocele. Autopsy findings showed right sided renal agenesis along with other anomalies detected by U/S.

There were 2 babies (Case No. 3 & 4) with dysplastic kidney disease due to early severe obstruction. In both cases obstruction was distal to urinary bladder, hence there was bilateral dysplasia of kidneys associated with dilated bladder.

Case No. 3: A still born male baby weighing 1.4 kg was born to 22 year old gravida 3, para 1 abortion 1, living 1 at 28 weeks of gestation. There was no h/o consanguinity. U/S report was hydronephrosis with gross dilatation of urinary bladder (common cloaca). Autopsy revealed, bilaterally enlarged kidneys, very much dilated urinary bladder, undescended testes and high ano rectal anomaly and urethral stenosis. All features were suggesting Prune belly syndrome.

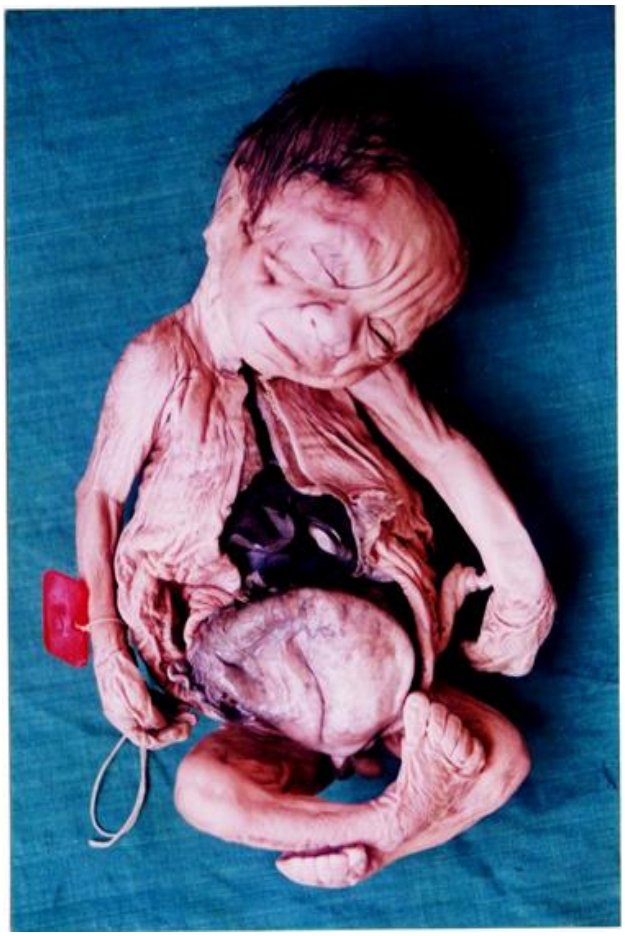

Figure 2: Prune belly syndrome: common cloaca.

Case No. 4: A still born male baby weighing 1.1 kg born to 22 year old gravida 2, para 1 at 26 weeks gestation to non-consanguineous couple. U/S report was severe oligoamnios, and dilated urinary bladder. Autopsy showed bilateral hydronephrosis and distended urinary bladder.

Figure 3 shows bilateral hydronephrosis with dilated urinary bladder.

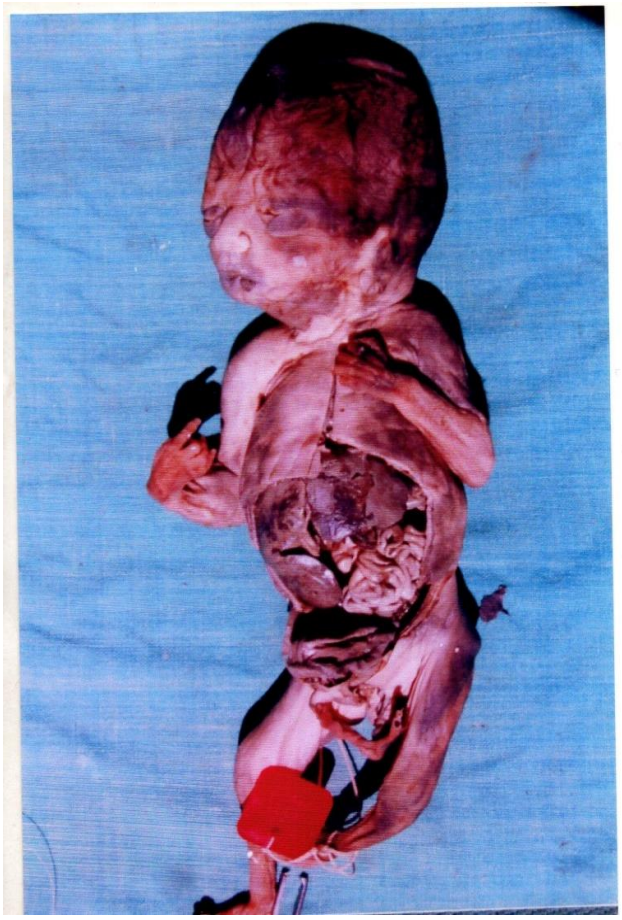

Figure 3: Bilateral hydronephrosis with dilated urinary bladder.

There was 1 live born male baby with bilateral hydronephrosis due to ureteropelvic obstruction.

Case No.5: A live male baby weighing 3.25 kg born to 32 year old gravida 2 para 1 at 40 weeks gestation to a non-consanguineous couple. Antenatal U/S showed mild oligoamnios and bilateral hydronephrosis. Post natal U/S showed bilateral hydronephrosis with minimal pelvicalyceal dilatation. Mother gives H/o similar findings in the 1st baby also suggesting recurrence of anomaly.

There was 1 live female baby with polycystic kidneys.

Case No. 6: A live female baby weighing 3.25 kg was born to 30 year old gravida 3, para 2 at 40 weeks gestation, antenatal U/S showed bilateral symmetrical enlargement of kidneys, bladder visualised and amniotic fluid volume was normal. Post natal U/S showed small cysts in echogenic kidneys. There was family history of bilateral polycystic kidney in the grandmother, mother and first baby.

Table 1 showing the details of cases observed in the present study based on ultrasonography.

Table 1: Showing results according to groups based U/S findings.

Group	Parity	Age	Gest age	CM	U/S report	Sex	Wt. kg	Live / stillborn	Autopsy reports
1	G1	19	32	0	Dilated lateral ventricle meningocele, omphalocele	F	1.75	SB	Omphalocele, meningocele, unilateral renal agenesis
	G2,P1,	21	32	2	Gross oligoamnios with IUD	M	1.25	IUD	Bil anophtalmia, bilateral renal agenesis, low anorectal anomaly, Fraser syndrome
2	NIL								
3	G3,P1, L1A1	22	28	0	Hydronephrosis, gross dilatation of urinary bladder	M	1.40	SB	Bilateral hydronephrosis, common cloaca, low anorectal anomaly, Prune belly syndrome
	G2P1 L1	22	26	2	Cystic lesion in pelvis, severe oligoamnios	M	1.1	SB	Bilateral hydronephrosis, distended bladder
4	G2P1 L1	32	40	0	Mild bil hydroneprosis with minimal pelvicalycial dilatation	M	3.25	Live	Confirmed after birth by fetal USG
	G3P2 L2	30	40	0	Symmetrically enlarged kidneys, bladder seen normal AFV	F	3.25	Live	Confirmed after birth USG echogenic cystic kidneys

DISCUSSION

Renal anomalies are grouped into 4 types based on antenatal ultrasonographic findings.[3]

Group 1 - Renal anomalies with associated major extra renal anomalies.

Group 2 - Renal malformation with non-visualisation of bladder showing poorly functioning kidneys. Group 3 - Bilateral renal malformation, oligoamnios and visualisation of bladder, with or without presence of cortico medullary differentiation.

Group 4 - unilateral or mild bilateral renal affection shown by cortico medullary differentiation with slightly reduced or normal amniotic fluid with visualisation of bladder. Termination of pregnancy was advised for group 1 & 2.

Table 1 showing the cases grouped as per this classification.

Incidence of renal anomalies in our study is 2/1000 births. The present study is compared with the studies reported in the literature.

Table 2: Showing comparison of incidence of renal anomalies with other studies.

Author	Total births	CA	%	Anomalies of US	Per 1000
A Takasande et al.[5]	9386	164	1.74	31	3.3
Neelam Grover et al.[7]	10100	180	1.78	7	0.69
Varsha Deshamukh et al.[8]	23843	124	0.52	13	0.54
Ohuka et al.[9]	8824	36	0.41	1	0.11
S. Swainet al.[10]	3932	48	1.2	6	1.52
Manish Kumar et al[3]	24160	422	1.74	63	2.60
Present study	3000	61	2.08	6	2.0

Bilateral renal agenesis is a lethal anomaly seen in 1 in 4000 births with male preponderance (2.5:1). Renal agenesis is due to failure of ureteric bud and failure of formation of nephrons and is usually associated with other system involvement[2]. The anomalies associated with renal parenchyma includes, renal agenesis,

polycystic kidneys, horseshoe kidney. There were 11 cases of renal agenesis in a study reported by Tulika Gupta et al.,[4] 8 cases in Manisha kumar et al.,[3] 4 in study reported by Amar T et al.,[5] 2 cases in the present study.

There are various reasons for renal cystic diseases, such as hereditary, developmental and acquired. Recent approach is to group the anomalies based on under lying cell biology & genetics.[2]

1) Early aberrant development - Dysplastic kidney with architectural distortion and cystic changes, due to failure of ureteric bud and induction of nephrogenic blastema.

2) Defects in terminal maturation - Polycystic kidney disease, where there is normal early development of nephrons and collecting ducts, later cystic dilatation of these structures causing secondary loss of adjacent structures.

Another practical classification of renal cystic diseases[2] - Dysplastic cysts - 1) Multi Cystic Dysplastic Kidneys (MCDK), 2) Dysplasia due to early severe obstruction and Hereditary cysts Polycystic kidney disease and inherited syndromes. 3) Non dysplastic, non-hereditary cysts - simple cysts. Malformed kidney is usually enlarged may be normal or small. There are multiple cysts of varying sizes that do not communicate with each other and are randomly distributed. Between the cysts is a dense stroma, but usually no normal renal parenchyma. Renal pelvis and ureter are atretic and are not visible. However in hydronephrosis, the dilated calyces are of uniform size, anatomically aligned, and communicate with the dilated renal pelvis.[2]

In the present study there were 2 babies (Case 3 & 4) with dysplastic kidneys due to early severe obstruction distal to urinary bladder. Manish Kumar et al.[3] reported 27 cases with dysplastic kidneys out of 55 babies with renal anomalies. T. Gupta et al.[4] reported 7 cases with pelvi ureteric obstruction among 34 renal anomalies.

Dilatation of upper urinary tract can be due to obstructive or non-obstructive-ureteropelvic junction obstruction - 44%, uretero vesical junction obstruction - 21%, vesico ureteral reflux - 14 %, posterior urethral valve - 9%.

Diagnosis of pyelectasis is made during antenatal U/S by measuring Renal Pelvis Diameter (RPD). If RPD measure ≥3 mm in 1st trimester, ≥ 4-5 mm in 2nd trimester, ≥ 7-10 mm in 3rd trimester suggest pyelectasis. Using RPD ≥4 mm at 18-23 weeks, and ≥10 mm at 28 weeks 96% of fetus with mild hydronephrosis resolve either antenatally or postnatally.[2] In the present study 1 live male baby (Case 5) had mild hydronephrosis due to obstruction at pelvi ureteric junction. And 1 Live female baby had polycystic kidney and history suggesting Autosomal Dominant Polycystic Kidney Disease (ADPKD). T. Gupta et al.[4] reported 8 cases with polycystic kidneys, Manish Kumar et al.[3] reported 16 babies of hydronephrosis due to distal obstruction.

Table 3: Comparison of types of renal anomalies with other studies.

Authors	Renal agenesis	Dysplasia of kidney	Distal obstruction	Other system involvement	Prune belly syndrome
T. Gupta et al.[4]	11	12	7	24	-
Manisha Kumar et al.[3]	8	27	16	13	3
Amir T et al.[5]	2	-	-	-	-
Present study	2	2	2	2	1

There were 2 babies with multiple organ involvement in the present study and Manish Kumar et al.[3] reported 13 babies with involvement other system.

CONCLUSION

Urinary system anomalies are common. Prognosis depends upon the type of renal anomaly and associated other system malformations. Assessment on prognosis of renal anomalies is made by amount of amniotic fluid, visualisation of urinary bladder and visualisation of normal renal parenchyma between cysts during antenatal ultrasound examination. Whenever renal pyelectasis is suspected repeat examination after 18 weeks and 28 weeks is done to confirm the renal anomaly as many times mild hydronephrosis resolve either before birth or after birth. Fetal autopsy after termination of pregnancy is essential to confirm findings and arrive to a definite diagnosis.

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Ethical approval: The study was approved by the institutional ethics committee

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