Unilateral Renal Agenesis Associated with Urinary Outflow Tract Obstruction in a Diabetic Pregnancy

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Abstract
We report a case of diabetic pregnancy where the foetus had enlarged cystic left kidney with agenesis of right kidney. The urinary bladder was dilated and urethra revealed posterior urethral valve an postmortem.

INTRODUCTION
Maternal diabetes is known to affect fetal organogenesis. Pre-gestational diabetes is strongly linked to major fetal malformations as hyperglycemia and its associated metabolic abnormalities lead to ‘fuel-mediated teratogenesis’. While cardiovascular and neurological malformations are more common and can be potentially lethal, other systems are also known to be affected. We report here a case of diabetic pregnancy with congenital anomalies involving kidneys and the urinary outflow tract.

CASE REPORT
A 33 year old Hindu married lady from a village in West Bengal presented for her first antenatal check up at 12 weeks of gestation. Although she was known to be diabetic for the last 2 years and hypertensive for the last 8 years, no information was available regarding her pre-conceptional glycemic control. Both her parents were diabetic. She was an oral antihyperglycemic agents at presentation and was never counseled on diet and exercise. Oral tablets were discontinued and the patient was advised insulin following which she achieved a reasonable degree of glycemic control, with fasting blood glucose in the range of 75-100 mg/dl and post-meal glucose levels in the range of 100-130 mg/dl. No other evaluation was done; no information on glycated hemoglobin levels prior to conception was available.

Her body weight was 63 kg and height was 152 cm. Blood pressure was 130/80 mm of Hg. No pedal edema was noted.

Blood examination showed hemoglobin: 10 gm%, fasting blood glucose: 75 mg/dl and postprandial blood glucose: 124 mg/dl, HbA1c: 7.5%, creatinine: 0.7 mg/dl, uric acid: 3.6 mgdl.

Ultrasonographic examination showed a gravid uterus with a single living fetus of 23 gestational weeks. A cystic ovoid structure was seen in the region of left kidney suggesting hydronephrosis; the urinary bladder was hugely distended, suggesting outflow tract obstruction.

When the patient and her husband were informed about the findings, they decided not to continue with the pregnancy. The pregnancy was terminated by ripening of cervix and prostaglandin E2 gel application; a dead male fetus weighting...
700 g was delivered. At autopsy, the left kidney was found to be enlarged and cystic (6 x 3 cm) while there was complete agenesis of the right kidney. The urinary bladder was distended and filled with urine (Fig. 1). Dissection of the urethra revealed posterior urethral valve.

**DISCUSSION**

Maternal diabetes leading to fetal malformations is known since long. The exact teratogenic mechanism in diabetes is not known. Hyperglycemia may be important but human studies focusing on the period of organogenesis are lacking. Hypoglycemia has also been suggested based mainly on animal experiments. Hyperinsulinemia appears likely. Numerous other factors including vascular disease, hypoxia, ketone and amino acid abnormalities, glycosylation of proteins or hormone imbalances could be also contributed to teratogenicity. The principal systems to be affected with fetus by uncontrolled diabetes in the mother, known as diabetic embryopathy are cardiovascular and neurological. Other systems affected are skeletal, e.g. caudal regression syndrome (pathognomonic of diabetic embryopathy), cleft lip and palate, arthrogryposis, etc; and gastrointestinal system, e.g. anorectal agenesis. Multiple malformations are common in embryo of a diabetic mother. The genitourinary system malformations are rare and present mostly as renal agenesis, ureteric duplication and pseudohermaphroditism.

The genitourinary system develops from urorectal septum; in a recent review² of urorectal septum malformation (URSMS), it was shown to consist of ambiguous genitalia concurrent with absence of perineal and anal openings. The sex ratio of the 13 new cases was seven males to six females and from the literature 21 males and 28 females. In addition, 11 of the 13 new cases had anorectal atresia with five of the cases also having partial agenesis of the colon. Bilateral renal agenesis was present in three of the 13 cases, unilateral renal agenesis occurred in six, and dysplastic kidneys were found in ten. The URSMS is a lethal condition with long-term survival reported in only three of a total of 62 new cases.²

Aphallia has been reported in a single case as part of diabetic embryopathy.³ Bladder outlet obstruction as seen in our case has not been reported previously. We believe this is the first reported case of bladder outlet obstruction as part of diabetic embryopathy.

**REFERENCES**


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**Announcement**

**International Symposium on Headache**

An “International Symposium on Headache” is being organized by the Department of Neurology, Chattrapati Shahruji Maharaj Medical University, Upgraded KG’s Medical College, Lucknow 226 003 on 1st and 2nd November, 2003 at Taj Residency, Lucknow. The faculty for the symposium includes Dr. M Ferrari (Netherlands), Dr. PR Saxena (Netherlands), Dr. PJ Goadsby (UK), Dr. NT Mathew (USA), Dr. SK Kackar (New Delhi), Dr. K Ravishanker (Mumbai), Dr. Ashok Chandra (Lucknow), Dr. Dipak Kumar (Lucknow), Dr. Rakesh Shukla (Lucknow) and Dr. RK Garg (Lucknow). Those desirous of participating should contact: Dr. Rakesh Shukla, Organising Secretary, Professor of Neurology, CSM Medical University, (Upgraded KG’s Medical College), Lucknow - 226 003, UP, India. E_mail : rakeshshukla_rakesh@rediffmail.com

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