

Case Report of Bilateral Renal Agenesis (Potter S Syndrome) at 26 Weeks Gestational Age

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ESID-5649-1369-202^b

Introduction

Bilateral renal agenesis or Potter's Syndrome is a very rare congenital anomaly presenting in one baby in 3000-4000 deliveries, it could have recessive autosomal etiology or sporadically. with a remarkable predominance of male infants(1, 2).

It might present with ('Potter's face') and pulmonary hypoplasia, the combination being known as Potter's syndrome. low set and malformed ears; long epicanthal folds; flattened bridge of the nose; small mandible; as well as renal agenesis, pulmonary hypoplasia, oligohydramnios, and amnion nodosum. There are frequent associated skeletal malformations such as clubbing of the hands and feet and other fixed and contracted joints. Similar facial, pulmonary, musculoskeletal, and amniotic findings have been described with a spectrum of renal abnormalities. The associations are reported by many researchers(1-4),the most common cause of oligohydramnios is renal agenesis or hypoplasia(5).In the course of pregnancy an increasing oligohydramnios becomes manifest; during labor, virtual absence of amniotic fluid is found in most cases. This oligohydramnios should alert the obstetrician to suspect Potter's syndrome, it is mentioned as a cause for developing the syndrome.(6) serial ultrasonography may confirm the diagnosis. premature delivery, and breech

presentation frequent finding. Consequently the prenatal diagnosis of BRA is important in order to avoid unnecessary operations(7)

Potter's syndrome" is often associated with the live-born but the infant can't be resuscitated, the cause of death is respiratory distress as a consequence of pulmonary hypoplasia.(1, 2)

Despite the remarkable facial characteristics of these infants, it was only in a small minority that the diagnosis was considered before autopsy. This stresses the need for a full post-mortem examination in all cases of perinatal death.

The multiple abnormalities of Potter's syndrome and of related syndromes suggest a chromosome defect. The etiology is still uncertain, though multifactorial inheritance is the most likely. As a consequence, the recurrence risk is not negligible; the small number of 'familial occurrence' observations, however, does not allow estimation of a risk figure. Genetic counseling is indicated in any family giving birth to a child with bilateral renal agenesis.

Some author reported sporadic case in which the mother consumed drugs during the 1st trimester.(3). Another reported a case of sirenomelia sequence with Potter's syndrome, the baby has absent urinary tract and external genitalia, the legs had separate bones and they were fused by skin and associated

with Potter's syndrome. His mother had gestational diabetes(8).

Differential Diagnosis

Bilateral Renal Agenesis (Mayer-Rokitansky-Kuster-Hauser Syndrome):

Multicystic Renal Dysplasia
Polycystic Kidney Disease
Posterior Urethral Valves
Prune Belly Syndrome
Sirenomelia
Ectopic Kidney:
Melnick-Fraser Syndrome
Fraser Syndrome

Prognosis

The outcome of Potter syndrome is poor. All the babies born are either stillborn or die very early(1, 2) Neonate associated with hypoplastic lungs are at very high risk, and the primary cause of death is due to respiratory distress syndrome within hours or days after birth. According to the experimental study conducted in John Hopkins hospital, regular saline injected in the mother's womb has enhanced the lung development. But dialysis is required throughout his/her life. A prognostic factor is determined by gestational age at diagnosis, type, and location if associated structural abnormalities. The survival rate is high in Potter syndrome with other causes rather than bilateral renal agenesis. The neonatal mortality rate is 100% if no obstetric interventions are done.

Complications

Renal insufficiency is the major complication of Potter syndrome. Major electrolytes imbalances such as hyponatremia, hypernatremia,

hyperkalemia, hypocalcemia, and hyperphosphatemia are due to renal failure. Obstructive uropathy favors urinary tract infections and hydronephrosis. Renal tubular dysplasia has many drawbacks likely erythropoietin deficient anemia, vitamin D insufficiency, and alteration of the renin-angiotensin system.

Most cases of Potter syndrome are associated with pulmonary hypoplasia. This ultimately leads to pulmonary insufficiency and respiratory distress syndrome. Such a neonate is cyanotic and suffers from respiratory acidosis due to the accumulation of CO₂ and ventilation-perfusion mismatch.

Physical structures deformities are the prominent characteristics associated with Potter syndrome. Insufficient amniotic fluid for proper fetal movement and body parts development, as well as uterine compression of the fetus, leads to deformities of the face and extremities. Genital abnormalities are present in up to 70% of cases. Other congenital heart defects, pancreatic cysts, esophageal atresia, duodenal abnormalities, colonic agenesis, Meckel diverticulum may be present. Subtype II is associated with hepatic fibrosis and biliary tree abnormalities. Multiple pregnancies are reported with intrauterine growth retardation, preterm labor, and preeclampsia.

Patient education

Potter syndrome is an autosomal inherited congenital disease with poor outcomes associated with multiple problems. Psychological effects in the family are common. So counseling of the family members, especially the mother about the consequences of the disease, plays a vital role in the management of the post-delivery stress(1)

Case report:

19 years old PG married for 1 year, her husband is her cousin. No family history of congenital anomalies, No significant past medical history nor drug history.

She as referred for anomaly scan, the pregnancy was uneventful till referral she was on good antenatal care received folic acid supplement.

Ultrasound findings:

- Single, viable, breech presentation.
- Posterior high placenta
- Anhydramnios
- GA by ultrasound 26+3days (HC:28+6, FL:24+3, AC: 24+4)
- Congenital anomalies:

- Hydrocephalus (severe ventriculomegaly measuring 22cm, normal is <10cm

Dilated third ventricle (6mm) thalami are not seen well.

- Both kidneys show features of multicystic changes

No cortico-medullary differentiation.

- Bladder filling is not seen

❖ Conclusion

- A picture suggestive of bilateral non-functioning kidneys.
- Hydrocephalus.
- No major anomaly seen however other anomalies can't be excluded due to poor visualization.

❖ **Impression**_very poor outcome.

Sonographic features

Note there was difficulty in taking these images as there is no liquor and even after amnioinfusion image quality is low

1. Fetal Biometry



BPD&HC



AC



Fetal viability



Coronal view of kidneys (Power Doppler study) (showed absent bilateral renal arteries) with absent fetal bladder



Coronal view of kidneys (showed absence of both kidneys)

Coronal view of kidneys (Color Doppler study) (showed absent bilateral renal arteries)



Antenatal intervention

Amnioinfusion (Before)

Note there is anhydramnios



Antenatal intervention

Amnioinfusion(After)

Deep vertical pool is 3.5cm



Report

Postnatal images

2D Measurements		AUA	Value	m1	m2	m3	Meth.	GP	Age
BPD (Hadlock)	✓		5.90 cm	6.01	5.78		avg.		24w1d
OFD (HC)			9.46 cm	9.20	9.71		avg.		
HC (Hadlock)	✓		24.90 cm	24.68	25.11		avg.		27w0d
HC* (Hadlock)			24.46 cm	24.18	24.75				26w4d
AC (Hadlock)	✓		18.84 cm	18.84			avg.		23w4d
FL (Hadlock)	✓		5.29 cm	5.29			avg.		28w1d

2D Calculations		Value	CI	GP
FL/BPD		90% (GA: OOR)		62% (70 - 86%)
FL/HC (Hadlock)		21% (GA: OOR)		28% (20 - 24%)
HC/AC (Campbell)		1.32 (GA: OOR)		

Postnatal images

Male fetus, 1.8 kg, delivered with signs of live in first of February 2018, at 18:00 pm, admitted to NICU due to severe apnea and died within one hour of delivery, there was no apparent congenital abnormalities





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International Journal of Applied Technology in Medical Sciences

Vol 1 Issue 1 (2022) 19 - 25

DOI: 10.54878

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International Journal of Applied Technology in Medical Sciences

Vol 1 Issue 1 (2022) 19 - 25

DOI: 10.54878

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