



POTTER SEQUENCE WITH IMPERFORATED IRIS- A RARE CASE REPORT

Dr. Bhargavi S	Junior Resident, Department Of Paediatrics, Sri Devaraj Urs Medical College, Sri Devaraj Urs Academy Of Higher Education And Research, Kolar.
Dr. Shrishail S	Assistant Professor, Department Of Paediatrics, Sri Devaraj Urs Medical College, Sri Devaraj Urs Academy Of Higher Education And Research, Kolar.
Dr. Gurram Kamalakara S	Senior Resident, Department Of Paediatrics, Sri Devaraj Urs Medical College, Sri Devaraj Urs Academy Of Higher Education And Research, Kolar.
Dr. K N Venkateshwara Prasad*	Professor, Department Of Paediatrics, Sri Devaraj Urs Medical College, Sri Devaraj Urs Academy Of Higher Education And Research, Kolar. *Corresponding Author

ABSTRACT

Introduction: Potter syndrome is a fatal congenital disorder characterized by the changes in physical appearances of neonate due to oligohydramnios caused by renal agenesis and impairment. **Case Report:** A Single live term female baby was extracted by LSCS with birth weight of 2. 50kg. Baby cried immediately after birth. Baby was shifted to NICU in view of Respiratory Distress. Antenatal scan done revealed Anhydramnios with AFI of zero. At admission to NICU baby was tachypnoeic and saturation being 80% on room air and 90% on HFNC support. Physical examination done revealed features of Porters sequence(wide open AF, cranial suture not fused, lowset ears, flat nasal bridge recessed chin, accessory nipple present, clenched hands with overlapping fingers, rocker bottom foot present), 30 min after admission respiratory distress worsened with desaturation in view of which baby was intubated and connected to Mechanical ventilator support chest Xray done revealed left sided pulmonary hypoplasia and ABG done revealed mixed acidosis, eye examination done revealed absent pupillary reflex. **Conclusion:** Potter's sequence is a fatal congenital disorder characterized by severe oligohydramnios due to renal anomalies, leading to pulmonary hypoplasia and distinct physical deformities. The prognosis is generally poor, with most affected infants not surviving beyond the neonatal period. Early prenatal detection is crucial for informed decision-making and supportive care

KEYWORDS : Potters, Anhydramnios, Imperforated iris.

INTRODUCTION:

Potter syndrome is a fatal congenital disorder characterized by the changes in physical appearances of neonate due to oligohydramnios caused by renal agenesis and impairment. It is incompatible with life as neonates with Potter syndrome have pulmonary hypoplasia that leads to respiratory distress within an hour of birth. Potter sequence and Potter syndrome are used interchangeably because the sequence of events leading to oligohydramnios is consistent. But the Potter sequence more specifically describes the decreased amniotic fluid irrespective of the cause.

Potter syndrome is classified on the basis of the cause of renal anomalies. Bilateral renal agenesis is classic to Potter syndrome. Subtype I is associated with autosomal recessive polycystic kidney, subtype II is due to renal dysplasia, subtype III is due to autosomal dominant polycystic kidney, and subtype IV is related with obstruction of ureter or pelvis causing hydronephrosis.¹

Case Report:

A Single live term female baby was extracted by LSCS with birth weight of 2. 50kg. Baby cried immediately after birth. Essential steps of neonatal care done. Baby was wrapped and shifted to NICU in view of Respiratory Distress. Antenatal scan done revealed Anhydramnios with AFI of zero. At admission to NICU baby was tachypnoeic and saturation being 80% on room air and 90% on HFNC support, systemic examination done revealed Bilateral sub costal and inter costal retractions with downes score of 4.

Physical examination done revealed features of Porters sequence(wide open AF, cranial suture not fused, lowset ears,

flat nasal bridge, recessed chin, accessory nipple present, clenched hands with overlapping fingers, rockerbottom foot present), 30 min after admission respiratory distress worsened with desaturation in view of which baby was intubated and connected to Mechanical ventilator support and UVC was secured, chest Xray done revealed left sided pulmonary hypoplasia and ABG done revealed mixed acidosis, eye examination done revealed absent pupillary reflex ophthalmologist opinion was taken and impression imperforated iris was made. baby was continued on MV support, Attenders were counselled regarding condition of the baby, explained about guarded prognosis, need for further evaluation and need for continuation of ICU stay and further plan of management but attenders were not willing for the same due to personal reasons, Hence discharged the baby against medical advice



Figure 1 Rocker bottom foot



Figure 2 Imperforated iris



Figure 3 Pulmonary hypoplasia



Figure 4 Baby with potters syndrome

DISCUSSION:

Potter's sequence describes the typical physical appearance caused by pressure in utero due to oligohydramnios. It can occur in conditions such as infantile polycystic kidney disease, renal hypoplasia, and obstructive uropathy. Some authors feel that Potter's sequence rather than Potter's syndrome is more appropriate terminology as all cases of this syndrome do not have exactly the same set of signs, but they share a common chain of events leading to the same endpoint of reduced or absent amniotic fluid. Decrease in the volume of amniotic fluid may be due to decreased urine production secondary to bilateral renal agenesis, obstruction to the urinary tract, or occasionally prolonged rupture of membranes. The resulting oligohydramnios is the cause of the deformities in Potter's sequence. Various abnormalities are associated with renal agenesis or dysplastic kidney abnormalities, such as renal coloboma syndrome, Kallmann syndrome, and branchiootorenal syndrome. Newborn male babies have an increased incidence of the Potter's sequence because they have a higher rate of Eagle-Barrett (Prune belly) syndrome and obstructive uropathy secondary to posterior urethral valves.

CONCLUSION:

Potter's sequence is a fatal congenital disorder characterized by severe oligohydramnios due to renal anomalies, leading to pulmonary hypoplasia and distinct physical deformities. The prognosis is generally poor, with most affected infants not surviving beyond the neonatal period. Early prenatal detection is crucial for informed decision-making and supportive care.

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