Potter Sequence

Potter sequence, also known as Potter syndrome, is a complication of oligohydramnios which results in characteristic clinical findings like limb deformities and craniofacial anomalies. The lungs fail to develop adequately (pulmonary hypoplasia) due to decreased amniotic fluid. Shortly after bith, the resultant neonatal respiratory distress is the major cause of mortality for these patients. Etiologies for oligohydramnios causing Potter sequence include autosomal recessive polycystic kidney disease (ARPKD), posterior urethral valves (PUV), chronic placental insufficiency, and bilateral renal agenesis.

PLAY PICMONIC

PATHOGENESIS

Oligohydramnios

Old-dry-uterus

Oligohydramnios is an amniotic fluid volume that is less than expected for the corresponding period of gestation. In the early weeks of pregnancy, the amniotic fluid is mostly water that comes from the mother's body. After about 20 weeks of pregnancy, the fetus's urine makes up most of the fluid. Low amniotic fluid volume can be caused by bilateral renal agenesis, obstruction of the urinary tract, polycystic kidney disease, or prolonged rupture of membranes. The resulting oligohydramnios causes the compression of the developing fetus in Potter sequence.

clinical findings

Limb Deformities

Voldemort-baby with Limb-deformities

Compression of the developing fetus leads to limb deformities like clubbed foot, hemivertebrae, and sacral agenesis.

Facial Anomalies

Voldemort's Facial Anomalies

Affected infants have facial anomalies such as low-set abnormal ears, retrognathia, flattened nose, and prominent epicanthal folds due to compression in utero.

Pulmonary Hypoplasia (Most Common Cause of Death)

Lungs and Hippo-plates with Death-Eater-Reaper

Compression of the chest and lack of amniotic fluid aspiration into fetal lungs leads to pulmonary hypoplasia. Its severity depends on the duration and degree of the oligohydramnios. Adequate space in utero is required for the movement of amniotic fluid into the fetal lungs and for the normal development of lungs. Hypoplasia is the most common cause of death in patients with Potter sequence due to respiratory failure soon after birth.

Etiologies

Autosomal Recessive Polycystic Kidney Disease (ARPKD)

Recessive-chocolate and Poly-parrot with Cystic-Kidneys

Autosomal recessive polycystic kidney disease (ARPKD) is characterized by cystic dilations of the renal collecting ducts. Due to the lack of urine creation and output because of renal abnormalities such as ARPKD, patients can develop oligohydramnios which then leads to the development of Potter sequence.

Posterior Urethral Valves

Post-terrier and U-wreath with Valve

Posterior urethral valves cause bladder obstruction, also known as obstructive uropathy. Patients with this anomaly have excess flaps of tissue in the posterior urethra which block the regular outflow of urine. Since urine cannot be voided from the bladder, it builds up within the kidneys in utero and can cause oligohydramnios, which can then lead to the development of Potter sequence.

Chronic Placental Insufficiency

Broken Placenta-present next to Old-crone

Chronic placental insufficiency, also known as uteroplacental vascular insufficiency, is a complication of pregnancy when the placenta cannot deliver an adequate supply of oxygen and nutrients to the fetus, and therefore, cannot support the developing fetus. It occurs when the placenta does not properly develop or when it has been damaged. Failure to deliver proper blood flow to the baby's kidneys can lead to decreased urine production, which in turn can cause oligohydramnions and lead to the development of Potter sequence.

Bilateral Renal Agenesis

Bi-ladders and A-apple-genie with Kidneys

Bilateral renal agenesis is a genetic disorder characterized by the absence or failure of both kidneys to develop in a fetus. Without proper functioning kidneys, the baby is unable to produce enough urine, which causes oligohydramnios, leading to the development of Potter sequence.