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Case Report of Prune Belly Syndrome

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Abstract

Introduction: Prune Belly syndrome/ Eagle Barrett syndrome. Incidence is 1 in 40,000 births. characterized by deficient abdominal wall muscles, undescended testes, urethral obstruction. majority of affected child born stillborn.

Case report: 26 yr. old female unbooked pregnant female. Gradvida 2, para2, abortion: -0, live birth: - 2. refered from Pvt hospital to new civil hospital with one day old child with Major Congenital Mal formation. baby was delivered at home.no TT, vitK was given to baby. baby was admitted in our NICU. on examination abdominal wall defect was present, ambiguous genitalia present, no anal and urethral opening present, undescended testis present. History taken from mother mother didn't have any antenatal visit nor taken TT n iron folic acid tablet. Patient was kept on O2 prong, IVF,1st line antibiotic. surgery reference done advised to refer to higher center.

Discussion: Exact etiology is not known. complication in perinatal period include oligohydramnios, Pulmonary hypoplasia, also cardiac anomaly, limb anomaly, anomalies of urethra, uterus and Vagina present. Prognosis depend on renal dysplasia and pulmonary hypoplasia.

Conclusion: Routine antenatal screening is must so that early detection of malformation can be made.

Keyword: Eagle- Barrett syndrome, undescended testis, limb Anomaly, anterior abdominal wall defect, ambiguous genitalia

Introduction

Also known as Eagle syndrome, Barrets syndrome, triad syndrome, Osler-parker syndrome, abdominal muscle deficiency syndrome. Incidence is 1in 40,000 live births. Males are affected more than females. Syndrome consists of: - a) deficient abdominal muscle, b) undescended testis, c) urinary tract abnormalities (severe urethral obstruction in fetal life).[1,2]

It is a rare genetic birth defect. Exact etiology is unknown.[3]

Fig 1:

Case Report

A 26yr old female G2P2L2A0 delivered a child with ambiguous genet ilia. Child born out of non-consanguineous marriage; baby was delivered at home with birth weight 2.3kg. baby delivered at 37 weeks of gestation, unbooked pregnancy, no iron/folic acid and TT injection taken during pregnancy, no antenatal USG done. Delivery conducted at home by dai, cord was cut with an old shaving blade CIAB, BW=2.3kg. O/E: - Tone=normal, HR=140/min, RR=50/min, RS=AE=BS, CVS=S1S2 normal, CNS=NAD, PA=Soft, CRTC <3sec, RBS=85mg/dl, SpO2=97%, Room air. on examination Length=47cm, CC=29cm, HC=36cm.

ambiguous genitalia was present, No anal opening present, bladder extrophy present, anterior abdominal wall defect, no urethral opening, CTEV present, testis were not palpable, multiple crease on abdomen.

Patient was kept on IVF, TT injection along with Tetanus immunoglobulin given. CXR, USGA+pelvis. Blood investigations done; 1st line antibiotics along with Inj VitK started. On 2nd day of life patient developed tachypnea O2 prong trial given, did not settle, so was shifted to HFNC. ABG and repeat investigations done.

HCO3 correction given, surgery reference done, ad viced to refer to higher centre but patients relative did not want to continue treatment hence took DAMA.

Fig 2:

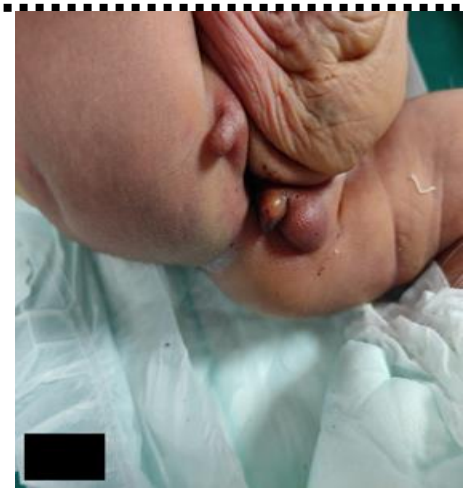

Fig 3:

Discussion

In 1839 frolin first described Prune Belly syndrome. Other association seen with Prune Belly syndrome include cardiac abnormalities, malrotation of bowel, musculoskeletal system defects or anomalies also present in urethra, uterus and vagina. Etiology of Prune Belly syndrome include embryological aberration of mesodermal development, fatal urethral abstruction, X-linked recessive chromosomal inheritance, oligohydramnios and delayed canalization.[4] It also shows VACTERAL association. Undescended testis needs to be fixed by 6 months of age by orchidopexy. For UTI antibiotics are required. Abdominal wall needs to be reconstructed, pulmonary hypoplasia and renal dysplasia determine the prognosis in Prune Belly Syndrome. Still birth and death is common.

Conclusion

Prune Belly syndrome is a rare condition. Surgery is required to fix the anterior abdominal wall defect, undescended testis, urinary tract problem.

There is no way to prevent the above condition. Routine screening for anomaly during pregnancy may help in early detection in country like India.

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