Proceedings of Virtual AOGD Monthly Clinical Meeting held at All India Institute of Medical Sciences, New Delhi on 25th June, 2021

Journey of Procreation in Cloacal Exstrophy: Defying all Odds

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Mrs X, 26-year-old woman $G_3P_{0+0+2+0}$ with 4 months amenorrhea presented to outpatient department for routine antenatal care. She was a known case of cloacal exstrophy with history of continuous dribbling of urine and involuntary passage of stools since birth. She was married for last 8 years. Her first two conceptions ended in first trimester missed abortions which were managed medically. Examination of respiratory, cardiac and neurological systems was unremarkable. Abdominal examination showed a complete absence of umbilicus and lower anterior abdominal wall. On local examination, the dorsal bladder mucosa was exposed with dribbling of urine seen from both ureteric orifices and urethral meatus was absent. Pubic rami were widely separated with a bifid clitoris. Labia majora were also widely separated. The distal part of rectum, anal canal and anal opening were exposed and there was no definitive anal sphincter. A small vaginal opening around 4 cm in length was present on extreme left side which was anteriorly placed and deviated towards the midline. A small cervix was felt at the tip of the finger. The vaginal opening on the right side was hypoplastic.

Obstetric examination revealed an enlarged uterus corresponding to 20 weeks of gestation deviated to left side. Foetal anomaly scan revealed a normal viable foetus corresponding to 19 weeks of gestation in left horn of the didelphic uterus. Routine investigations revealed moderate anaemia which was corrected by injectable ferric carboxymaltose as she was intolerant to oral therapy. Ultrasound abdomen showed normally situated left kidney with mild hydroureteronephrosis and right pelvic kidney. At 33 weeks' gestation, she went into spontaneous preterm labour. Antenatal steroids (dexamethasone 6 mg 12 hourly) and tocolysis with nifedipine were administered but she did not respond. Emergency caesarean section was performed in view of malformed pelvis and breech presentation. Prior to caesarean section, both ureteric orifices were identified and catheterized by infant feeding tubes. The abdomen was opened by left paramedian vertical incision keeping short of the exposed bladder. The rectus sheath and muscle were very thin and deficient and the left gravid horn was completely shifted to the left. A vertical incision was made in the upper part of the lower segment and a healthy male baby weighing 1602g was delivered as breech. Baby needed ICU care for 72 hours. Both baby and mother were discharged in good condition after 3 weeks.

Discussion: Cloacal exstrophy is a rare complex congenital birth defect which occurs due to abnormal development of the cloacal membrane during the embryonic period. It is the most severe form of the group of anomalies called bladder-exstrophy–epispadias complex (BEEC), as the lower abdominal wall is congenitally deficient. It occurs in approximately 1 in 200,000 to 1 in 400,000 live births with a male predominance. As reproductive function is preserved pregnancy is reported in previously operated cases. Only 5 cases of successful pregnancies

have been reported in women with corrected cloacal exstrophy. To the best of our knowledge this is the first report of a successful pregnancy in an uncorrected case of cloacal exstrophy.

Foetal intervention with international Collaboration, Administrative Support and Social Services to the Rescue of an Unborn foetus

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The Rh blood group system in humans is known to cause haemolytic disease of foetus and newborn (HDFN). We report a rare case (1st reported case in India) of successful outcome of HDFN due to anti Rh 17 in a woman with D - - phenotype which is characterized by the absence of C, c, E, e antigens and overexpression of D antigens. Globally, previously there are only 18 reported cases of Rh D-- in pregnancy, out of which, 8 had successful outcomes.

She is 27 year old female, $G_8P_4L_0A_3$ with bad obstetric history of previous 4 intra-uterine demises between 28-36 weeks and history of eclampsia in one pregnancy. She was diagnosed with this rare blood group during her 6th pregnancy at 23 weeks period of gestation when she developed foetal hydrops and severe maternal anaemia. Her blood was incompatible with all blood groups. At that time she was referred to our institution and workup for rare blood groups (Bristol laboratory, UK) confirmed this Anti Rh 17(D--) status. Then she received plasmapheresis and delivered a macerated hydropic baby.

In current pregnancy, patient reported at 19 weeks with foetal anaemia. Initially she received 2 cycles of intravenous immunoglobulin but despite that the foetus developed hydrops at 21 weeks. Procuring this rare blood for intrauterine transfusion (IUT) was the biggest challenge for which international rare donor panel was contacted. Nearest compatible donors were identified in Japan. Even though blood was available free of cost, each instalment of transport cost was around 2000 USD. With the help of hospital administration and due diligence from the medical social officers, funds were arranged. Appropriate import permits were taken.

The patient received 6 IUT's, first at 21+6 weeks and the 6th at 28+1 weeks. Steroid cover was given at 28 weeks. At 31 weeks, the foetus developed non-reassuring non-stress test and a preterm caesarean section was done. Preoperative autologous blood transfusion was performed for the patient due to non-availability of blood for the mother. The baby received surfactant, 1 cycle of partial exchange transfusion and 6 cycles of phototherapy. Baby was discharged in stable condition day 29 of birth.

This case was successfully managed with a close collaboration between foetal medicine, transfusion medicine and the neonatology team. Establishing a rare blood group registry in developing nations can help in managing such cases with relative ease.

A Case of ITP Gone Rogue in Pregnancy: Challenge Embraced

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Refractory immune thrombocytopenia (ITP) in pregnancy is clinically challenging for obstetricians and haematologists. First-line treatment includes corticosteroids, intravenous immunoglobulin (IVIG). Novel drugs, Romiplostim & Eltrombopag (thrombopoietin receptor agonist) which form second line treatment do not have well controlled trials in

pregnancy and have been rarely used. Splenectomy is usually kept as the last resort and is preferably done in second trimester. We report a rare case of refractory ITP in pregnancy where sequential pharmacological management including novel agents followed by splenectomy were used.

A 30-year-woman, $G_5P_1L_1A_3$, with diamniotic dichorionic twins at 24+3 weeks gestation, Rh negative (non-immunized) was diagnosed with severe ITP. Medical management including high dose steroids and IVIG could only cause transient improvement. Her platelets remained in the range of 5000-10,000/cumm. In view of limited evidence, after multidisciplinary deliberations and patient counselling, a trial of novel agents like TPO receptor agonist was given after counselling. She received multiple platelets transfusion (98 RDPs and 1 SDP). At 35 weeks, in view of FGR and poor biophysical profile LSCS along with splenectomy was performed. The intra-operative and post-operative course was uneventful and both babies had no evidence of thrombocytopenia. Maternal Platelet counts again dropped to 30,000/mm^3 on post op day-4 and oral Eltrombopag was tried which showed a favourable response. She was discharged in stable condition with platelet count between 40-50,000/mm^3 on post-operative day 15.

Splenectomy in pregnancy or during caesarean section is rarely reported. From our institution, Mahey et al (2013) have reported similar case where splenectomy was conducted at 38 weeks along with caesarean section.