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PRUNE - BELLY SYNDROME- A CASE REPORT

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Baby of Khina Maya Magar, full term, male a product of nonconsanguinous marriage was born to a 34 years old 3rd gravida mother with previous history of one male neonatal death (after 8 hrs) and one live female child. This was an unbooked pregnancy with no antenatal check-ups. She came in 3rd stage of labour with thick meconium stained liquor and delivered a baby with poor Apgar score of <3 at 1 minute, needing all resuscitative measure including IPPV and emergency resuscitative medications.

On examination baby was full term, male, weighing3.4 kg. He had facial dysmorphism s/o potter facies, and largely distended abdomen with massive ascites and bilateral palpable renal lumps measuring 12x6 cm. Baby also had midline cleft palate. Respiratory system examinations revealed bilateral crepitations. Rest of the systemic examination were within normal limit.

Initial investigations included septic screening which was within normal limit. There was right sided pneumo- thorax with collapse of left lung and shift of mediastinum to the left. On ultrasound abdomen showed bilateral hydronephortic kidneys with magaloureters and distentded urinary baldder. There was evidence of free fluid in the adbomen.

With the above clinical findings, a diagnosis of Prune - belly syndrome was put forward. Retrospectively, relevant maternal history was taken, which showed mother was a chronic smoker. There was no maternal history suggestive of any medication during 1st and 2nd trimesters. There was history of one previous male neonatal death with similar course.

Baby had neonatal respiratory distress syndrome. He was put on bag and tube ventilation but expired at 12 hour of life despite the best resuscitative efforts.

DISCUSSION

Prune- belly syndrome is a rare neonatal emergency seen in one in 40,000 deliveries, almost invariably in male. There may be maternal history of polyhydramnios. Baby usually has potter facies, obstructive uropathy usually due to PUV (Post urethral valves) with bilateral dysplastic kidneys and genital abnormalities in the form of elongated phallus. Typically there is congenital deficiency of adbominal musculature, dilatation of urinary collecting system and bilateral cryptorchidism with testes usually intra-abdominal. Malformation of bone, cardio-vascular system and gastro instestinal tract (malrotation) may also be involved. Babies are usually born limp and

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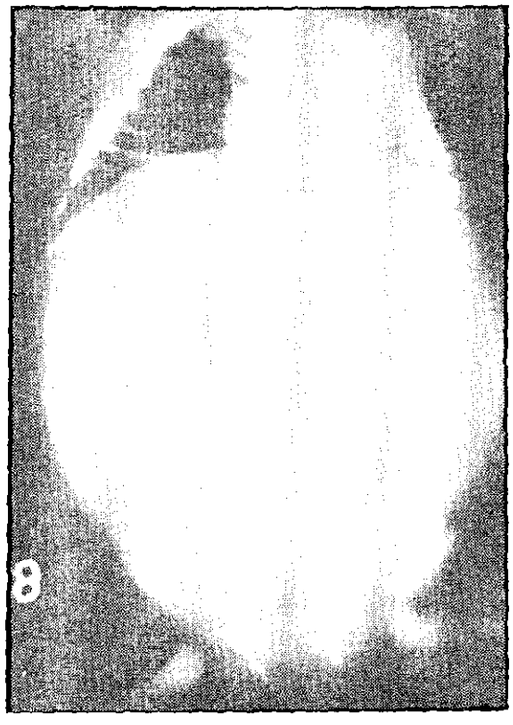

X-ray showing- Rt sided Pneumathorax, collapsed if lung & B/L Hydronephrotic Kidneys evidence of free fluid in abdomen.

blue needing all resuscitative meaures. With vigorous resuscitative efforts they may land up with pneumothoraces. Many are still born. Even in level III care neonatal centres mortality ranges any where between 90-95% and death is usually due to pulmonary hypoplasia and ventilatory problems as it occured in our case. Survivors will have chronic renal failure, needing dialyses and renal transplant.

Treatment modalities avaliable are fetal therapy which includes antenatal detection and placement of vesico-amniotic shunts and fulguration of PUV by fetal surgeons or termination of pregnancy if detected earlier in 1st trimester which is not possible in our set up and so the mother was advised genetic counselling before another pregnancy.

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Dear Dr Joshi,

The previous case report on [illegible] was published in Jan-Mar 1998 issue. Please refer it and revise your article accordingly.

Waiting for your prompt response.

Sincerely,

Ramesh