

Umbilical abnormalities

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Common symptoms

Common symptoms

- Mass
- Discharge
- Umbilical hernia
- infection

Umbilical mass

Umbilical mass

- Umbilical granuloma –
- Umbilical polyp

Umbilical mass

- Umbilical granuloma –
no associated anomalies
respond to 75%AgNO₃
or excision
- Umbilical polyp
surface lined by gastric or
intestinal mucosa
30-60% chance of another OMD
anomaly

Other rare masses

Other rare masses

- Urachal cyst
- OMD cyst
- Dermoid cyst
- Teratoma
- Rhabdomyosarcoma
- Fibrous histiocyteoma

Umbilical discharge

Umbilical discharge

- Scanty- UG or Polyp
infection
Eczema
- Copious – OMD or Urachal
remnant
- Pilonidal disease

Omphalitis

- Rare now
- Can progress quickly to necrotising fasciitis and portal vein thrombosis.

Umbilical Hernia

- Incidence :
- Higher incidence

Complications are extremely rare

Umbilical Hernia

- Incidence : 10-20% of all infants
75% of < 1500 g
- Higher incidence : African descent
Beckwith –W s
Downs

Complications are extremely rare

How do you manage UH

Mx of UH

- DD < 0.5 cm – 96% close spontae
> 1.5 cm unlikely to close
- Usual practice is to watch for 4-5yr
unless symptomatic or large
defect

Read more on

- MD
- OMD
- Urachal remnants / Ca

Congenital abdominal wall defects

Historical Background

- 1634 - Pare' Earliest recorded description .
- 1803 - Hey & Hamilton 1st successful primary repair.
- 1887 - Olshausen described mobilization of skin flaps.
- 1899 - Ahlfeld described Escharotic treatment using alcohol.
- 1956 - Cunningham popularized it.
- 1957 - Grob used 2% Mercurochrome.

Pare's original description of omph

LIB. 24.

Concerning the Generation of Man.

959

CHAP. LXVI.

Of the relaxation of the navell in children.



Fret times in children newly borne, the navell swelleth as bigge as an egg, because it hath not bin well cut or bound, or because the whayish humours are flowed thither, or because that part hath extended it selfe too much by crying, by reason of the paines of the fretting of the childe guts, many times the childe bringeth that tumour joined with an absesse with him from his mother wombe: but let not the Chirurgian assay to open that absesse, for if it be opened, the guts come out through the incision, as I have seene in many, and especially in a child of my Lord *Matrigues*; for when *Peter* of the *Rocke*, the Chirurgian, opened an absesse that was in it, the bowels ranne out at the incision, and the infant died; and it wanted but little that the Gentlemen of my Lords retinue that were there, had strangled the Chirurgian. Therefore when *John Gromontius* the Carver desired me, and requested mee of late that I would doe the like in his sonne, I refused to doe it, because it was in danger of its life by it already, and in three daies after the absesse broke, and the bowells gushed out, and the childe died.

An absesse no
to be opened

A histour

Figure 68.1 Pare's original description of omphalocele.

Classification

Classification

- Omphalocele
- Gastroschisis
- Bladder Extrophy
- Cloacal Extrophy



Omphalocele



What is this



Gastroschisis



What is the pathology



Hypogastric omph & cloacal extrophy

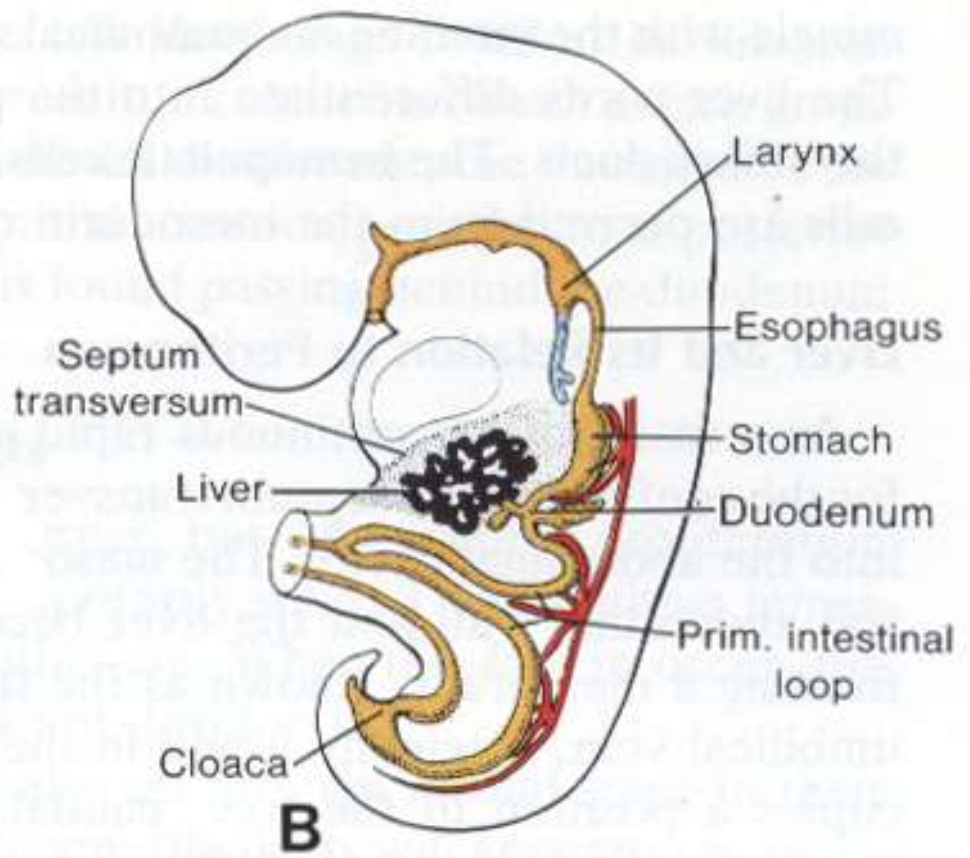
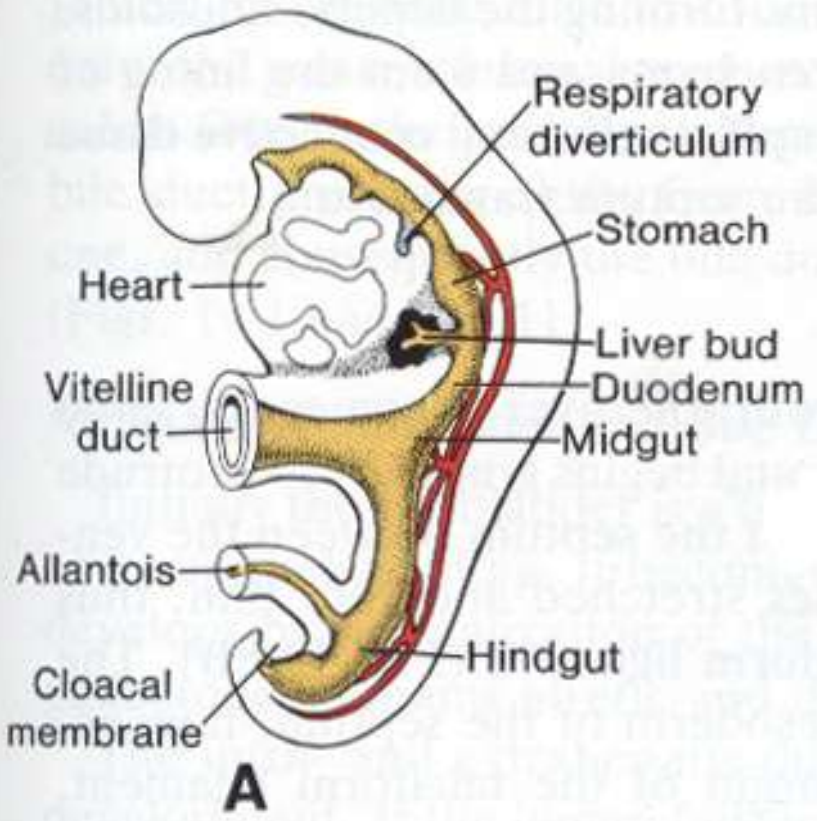


Embryology

- During 6th week of fetal life intestines grow rapidly and migrate out of the umbilical ring into the umbilical cord.
- During 10th week it returns back to the abdominal cavity rotating 270⁰ counter clockwise.

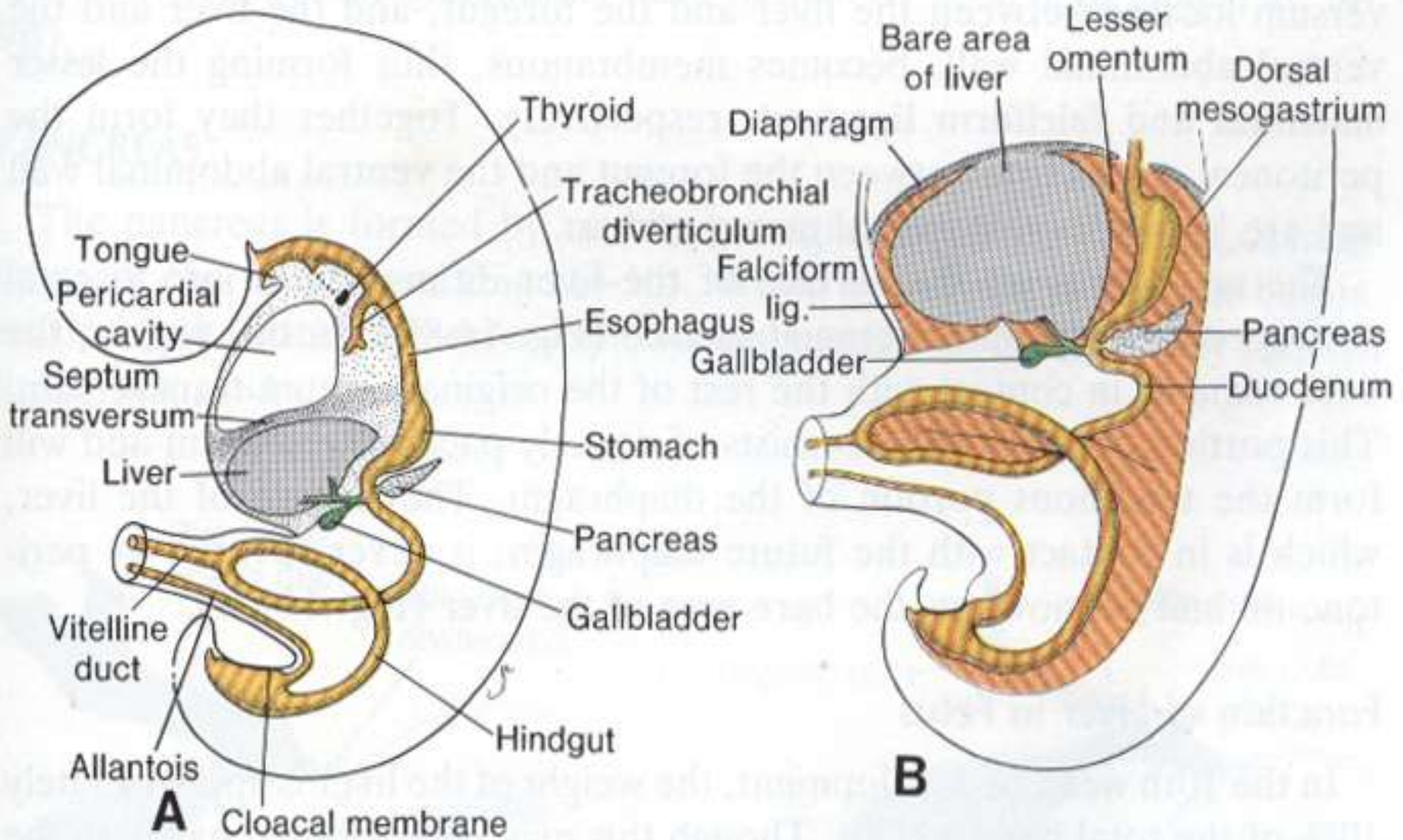
Embryological Basis

- Omphalocele- Failure of the midgut to return due to delayed closure of the lateral folds resulting in a large umbilical ring.
- - Failure of the cephalic folds results in Pentalogy of Cantrell
- -Failure of the caudal folds to close results in a hypogastric omphalocele with bladder or cloacal extrophy

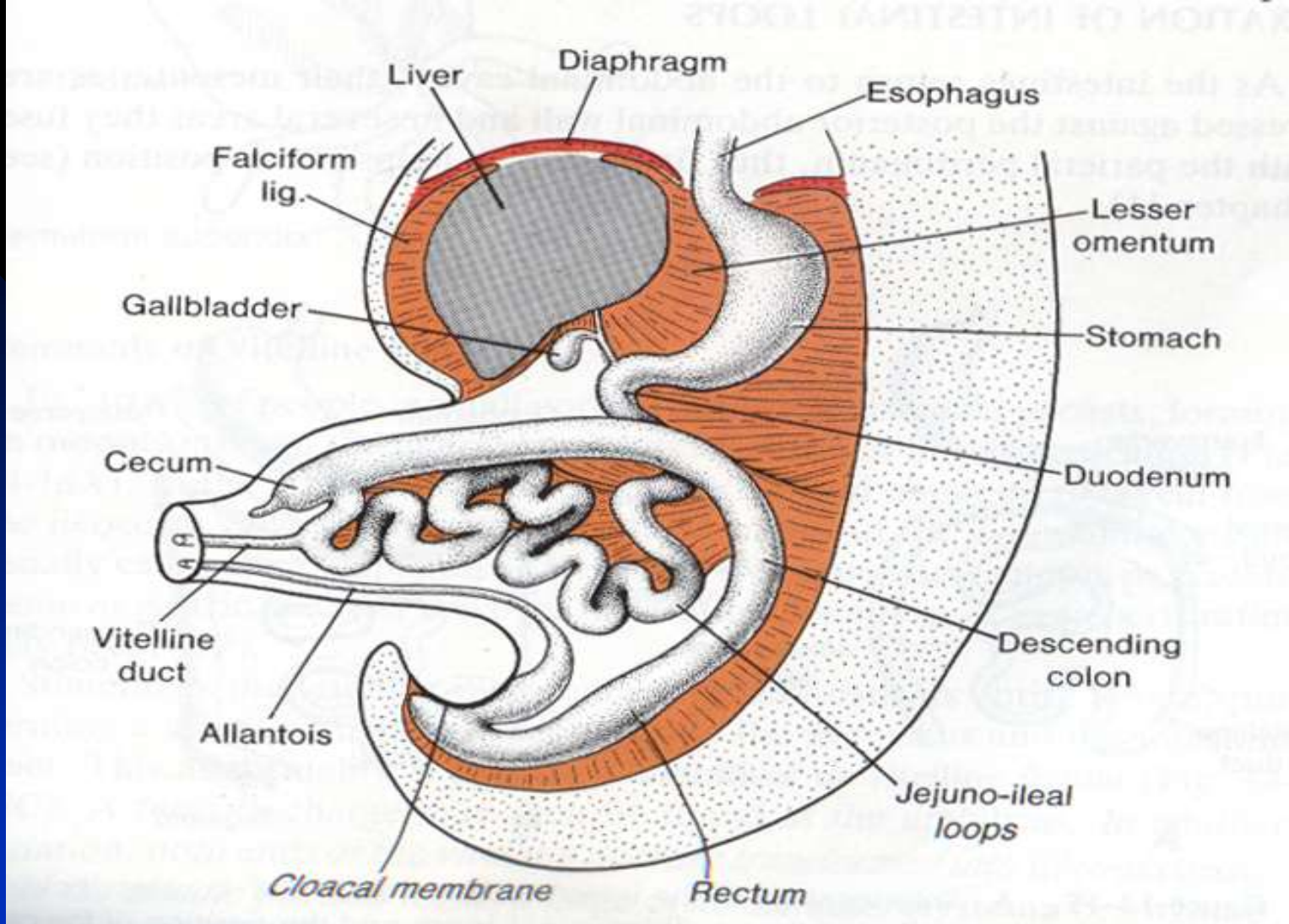


A . 25 day embryo – Short primitive midgut

B. 32 day embryo – Formation of primitive intestinal loop



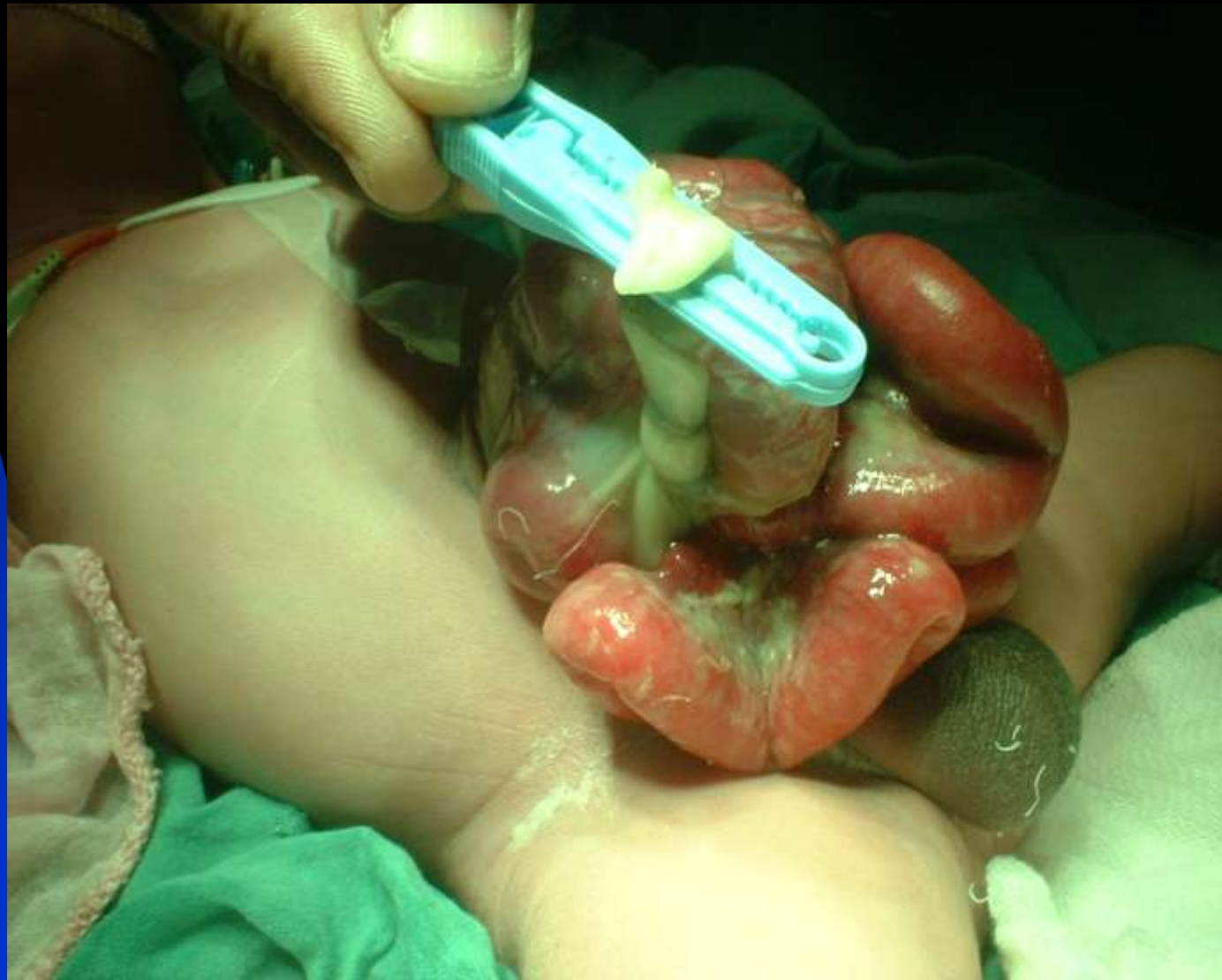
36 day embryo – liver expands caudally & midgut grows rapidly



8 week embryo – Physiological umbilical herniation of bowel loops

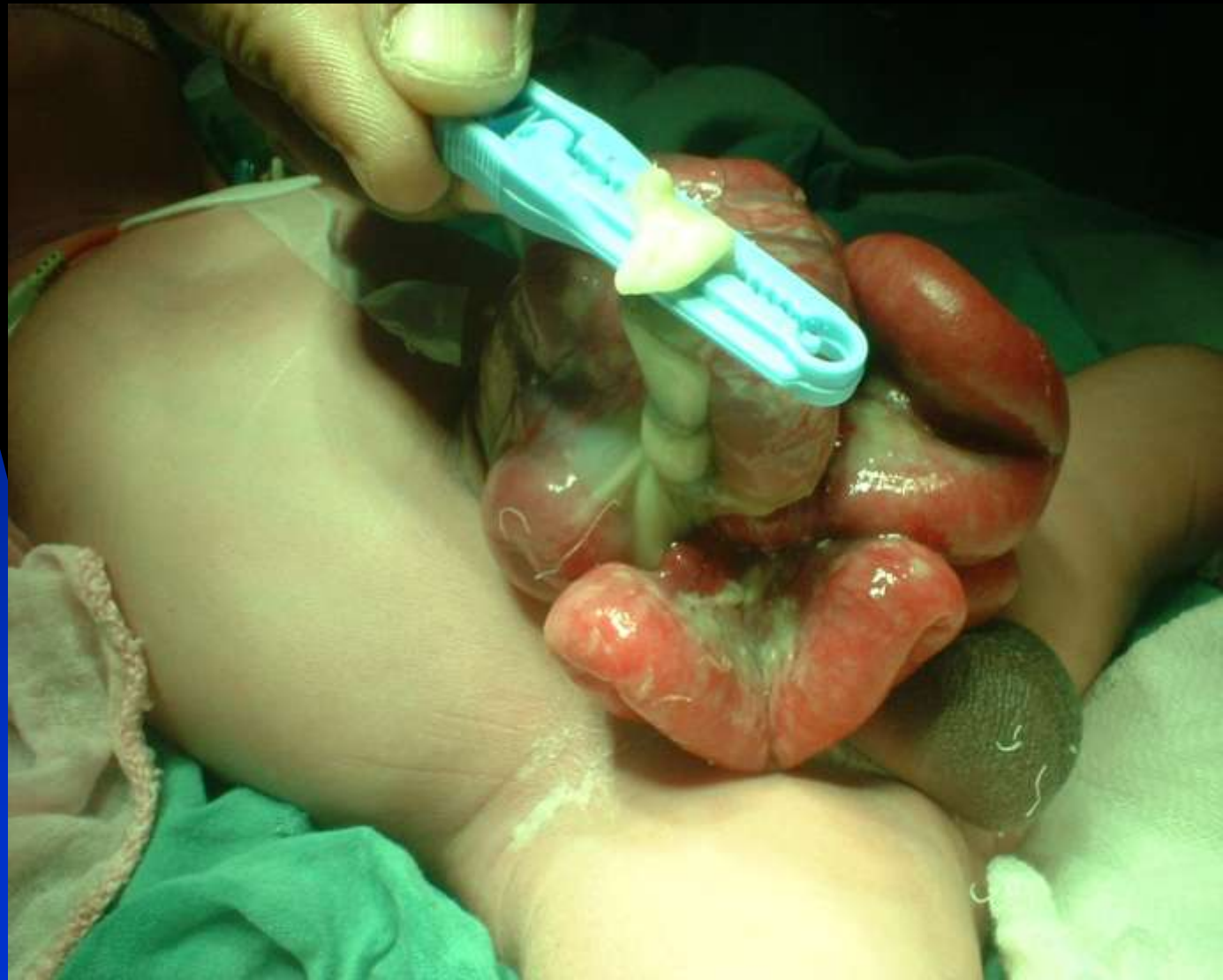
Embryo of Gastroschisis

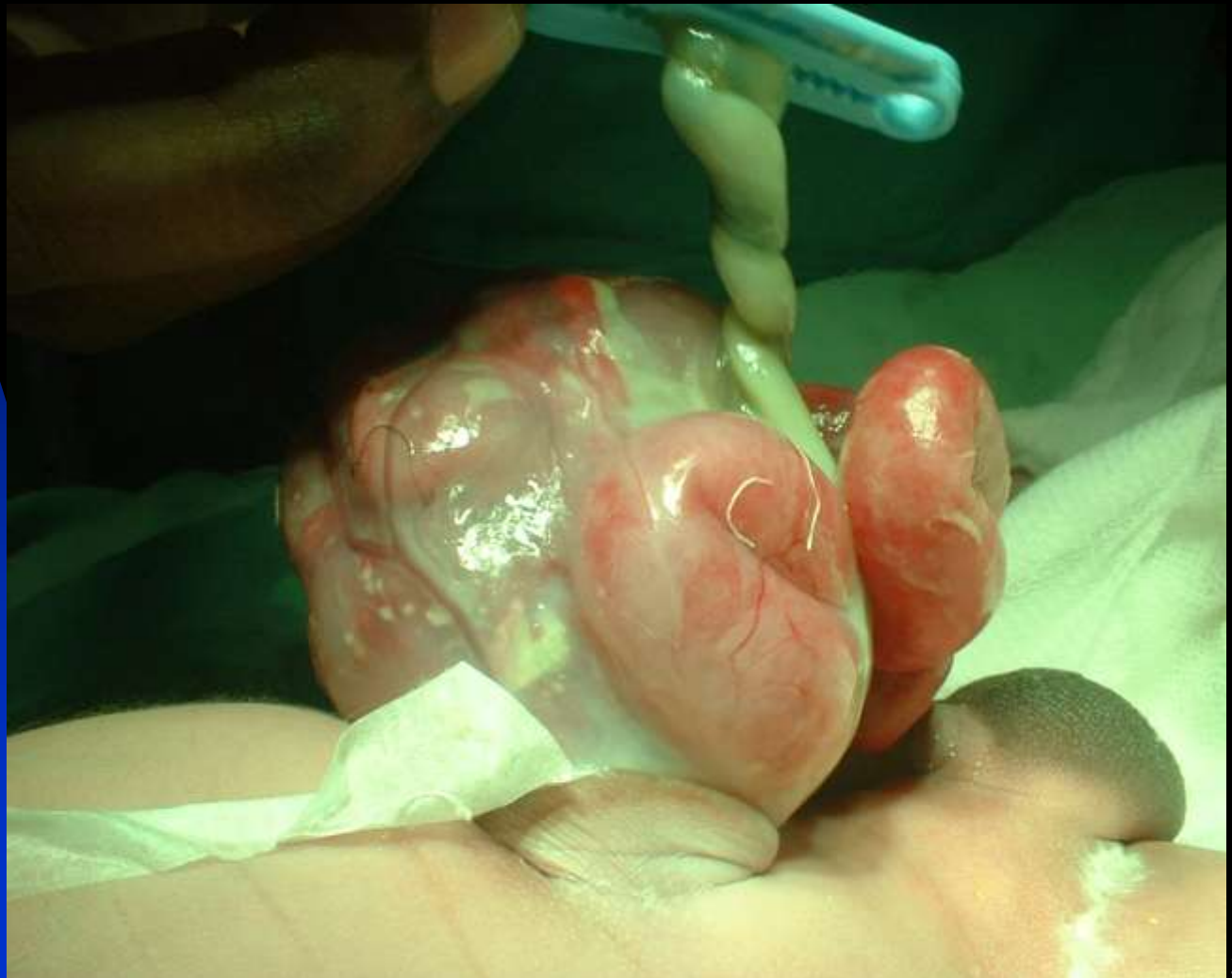
- A vascular accident involving right omphalomesenteric artery.
- A ruptured omphalocele in utero.



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Ruptured Exomphalos





Omphalocele ruptured at birth



Incidence

- Omphalocele
 - 1 in 4000 LBs
 - Remains the same over the years
- Gastroschisis
 - 1 in 6000 to 10000 LBs
 - increasing over the last 30 yrs .
 - Commoner in teen pregnancies.
- In USA combined incidence 1 in 2000₃₈

Risk factors for Gastroschisis

- Teenage mum
- Smoking
- Recreational drugs
- Low BMI
- GU infections

Incidence at LRH

- All babies with Omp or Gas admitted to LRH From 30.6.2005 to 1.7.2006

Omphalocele – 26

Gastroschisis - 6

Total births in Sri Lanka 365,145.

(Crude BR 19.1 / 1000 pop,

Mid year pop – 18,955,000)

- Estimated no of AWDs if incidence is similar to the West – 180/ yr
- The No of AWDs operated by Paed surgeons in Sri Lanka – 38 /yr
- Inference : Incidence of AWD is v low in Sri Lanka or majority of them do not reach a PSU. ie. Operated by general surgeons / dies before reaching a PSU.

Associated Anomalies



Associated Anomalies

- Omphalocele – 72%
 - Cardiac : 20% ASD, TOF
 - Trisomies : 20% Tri 13, 14, 15,
18 & 21
 - Beckwith- Wiedemann Synd : 12%
 - Pentalogy of Cantrell
 - Lower midline synd

Associated Anomalies

- Gastroschisis-

Other anomalies rare.

GIT anomalies : Atresias (10-15%)

: Meckels Diverticulum

: Intestinal Duplications

The function of exteriorized viscera

- Omphalocele – Normal because of membrane.
- Gastroschisis – Intestinal damage from exposure to amniotic fluid (due to meconium than urine in AF)
 - Constriction at the abdominal wall defect.
 - Changes in collagen composition & production of mucosal enzymes.
 - Raised inflammatory cytokine levels in amniotic fluid.

GASTROSCHISIS

- Intestinal transit and absorption returns to normal within 6 months in most babies

Prenatal Diagnosis and Management

- Cannot diagnose before 10th week as it is normal to have viscera outside the abdomen.
- Maternal serum AFP is raised in
 - Omp : 90%
 - Gas : 100%
- With both Ultrasound and MSAFP the AN diagnosis approaches 100%

What Next

- Look for associated anomalies
- Gas : Only careful anatomic U/S
- Omp : Structural defects
 - Anatomic U/S
 - Fetal Echocardiogram
- : Chromosomal defects
 - Amniocentesis
 - Chorionic villus sampling

Ante natal interventions

- Omp : No major role
- Gas : Amniotic fluid exchange or amnioinfusion in those with oligohydroamnios .

Timing of Delivery

Timing of Delivery

- Omp : Term delivery.
- Gas : Early delivery reduces the duration of exposure to AF.
37th week is a good time
(controversial)

Mode of Delivery

- Omp : V. large sac with herniated Liver > Caesarean del.
: Small sac > NVD
- Gas : No advantage in Caesarean except early delivery.

Location of Delivery

- All deliveries should be at a perinatal centre with neonatal surgical expertise.

Management – Newborn

- Initial care :
 - * Fluid resuscitation – need 2-3 times the normal req.
 - * NG decompression
 - * Avoid hypothermia.
 - * Cover the exteriorized viscera – warm saline soaked towel with a water proof dressing around.

Initial care – Gastroschisis.

- Bowel should be inspected to ensure adequate blood supply is not compromised by twisting of the mesentery or constriction at the wall defect.
- If size of defect is too narrow causing vascular compromise enlarge it immediately.

Surgical Management

- Goal – Place herniated viscera back in the abdomen and close fascia & skin.
- Strategies to facilitate above:
 - Stretching of abdo. Wall
 - using saline or mucomyst enemas to clear meconium from the colon.
 - Milking the small bowel contents toward stomach & aspirating with NG tube.

(cont.)

- Before replacing bowel
 - Look for : Atresias
 - : Malrotation / Non rotation
 - : Defects in diaphragm.
 - Arrange bowel in a position of non-rotation.
- If intra abdominal pressure too high > silastic silo > gradual reduction of contents over the next 1-10 days (every 12-24 hrs)

Repair of Gastroschisis

- Immediate repair in delivery room
(**Detroit group**)
 - Easier abdominal wall closure.
 - Earlier extubation
 - Early oral feeding
 - Shorter hospital stay.
- Routine bedside placement of a **spring loaded silo.**
- **Bianchi – Minimal intervention management** (Elective delayed reduction without GA)











Abdominal Compartment Syndrome

- Due to raised intra-abdominal pressure reduced perfusion of
 - GIT > NEC , Perforation
 - Liver > Hepatic failure
 - Kidney > Renal failure
 - Lower extremities > congestion & oedema

VAD and IAH

- Viscero abdo disproportion
- IAH : > 15 mmHg
- ACS ; > 20 mmHg

How to monitor intra abdominal pressure

- Peak Airway pressure < 25mm Hg
- Intra gastric / vesical / rectal pressure - < 20 mmHg
- CVP < 4 mmHg.

Post op management

- Feeding – Delayed in Gastroschisis
(may take weeks to months)
 - Prokinetic agents cisapride
- Almost all need assisted ventilation
 - Improvements in methods of neonatal ventilation and advances in neonatal TPN is responsible for the improved outcome in recent times**
- NEC : common in Gastroschisis
- GOR : Increased incidence

Escharotic Mx of large Omphalocele

- In Giant Omphaloceles the sac is left intact and allowed to granulate & eventually epithelialise.
- Silver Sulphadiazine is applied on the sac to prevent bacterial colonisation. (mercurochrome / iodine is no longer used due to reports of toxicity)
- The resulting ventral hernia is electively repaired when the risks are low

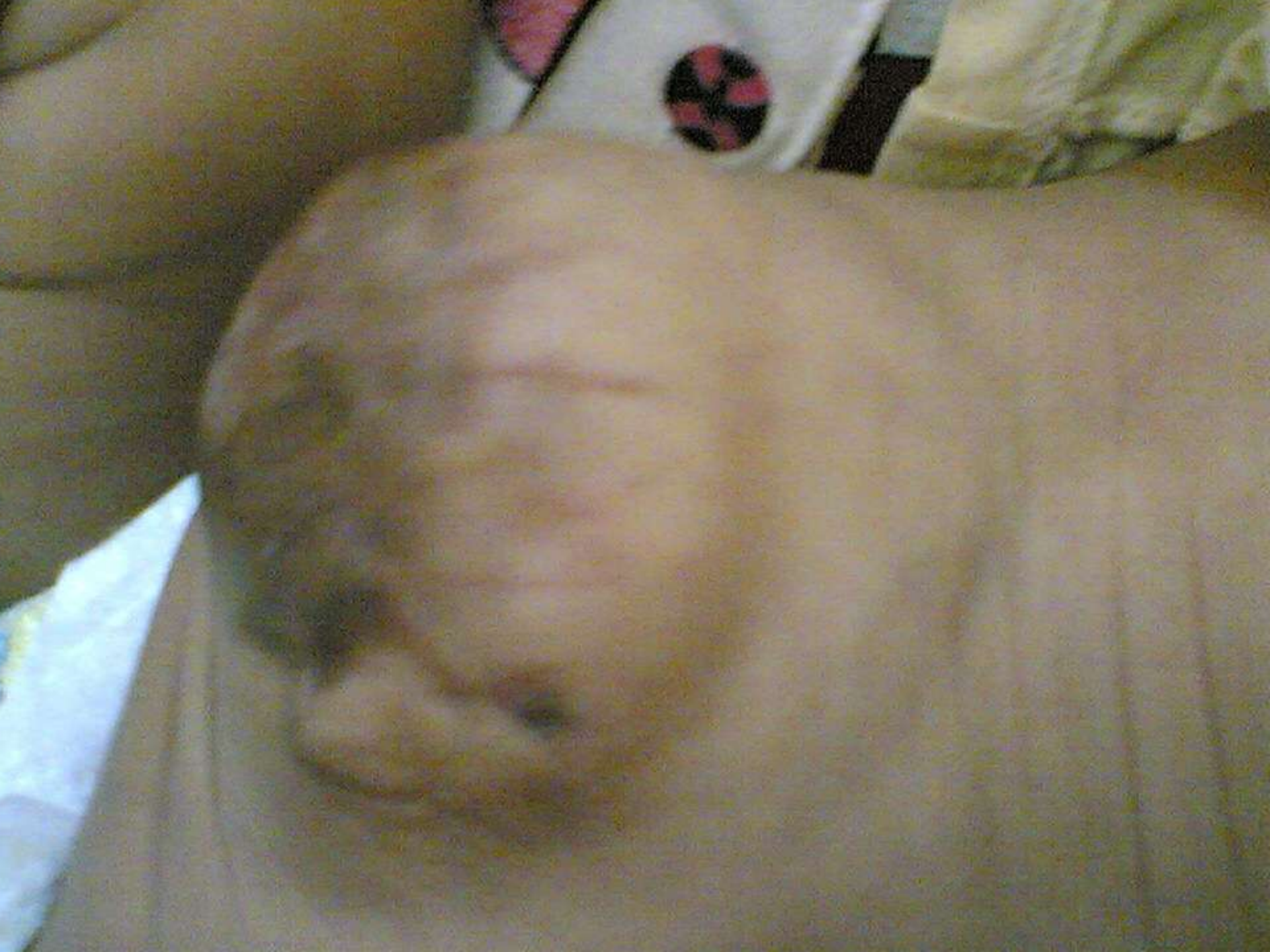




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Large ventral hernia





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Post op







Long term outcome

- Gastroschisis – Excellent in 90% of the cases

Problems : Adhesive obstruction

: Short bowel syndrome

: Motility disorders

- Omphalocele – Depends on the associated anomalies.

Excellent outcome in the absence of other anomalies.

Thank you