Esophageal Atresia

Long Island College Hospital
Daniel H. Hunt, M.D.
May 5th, 2006

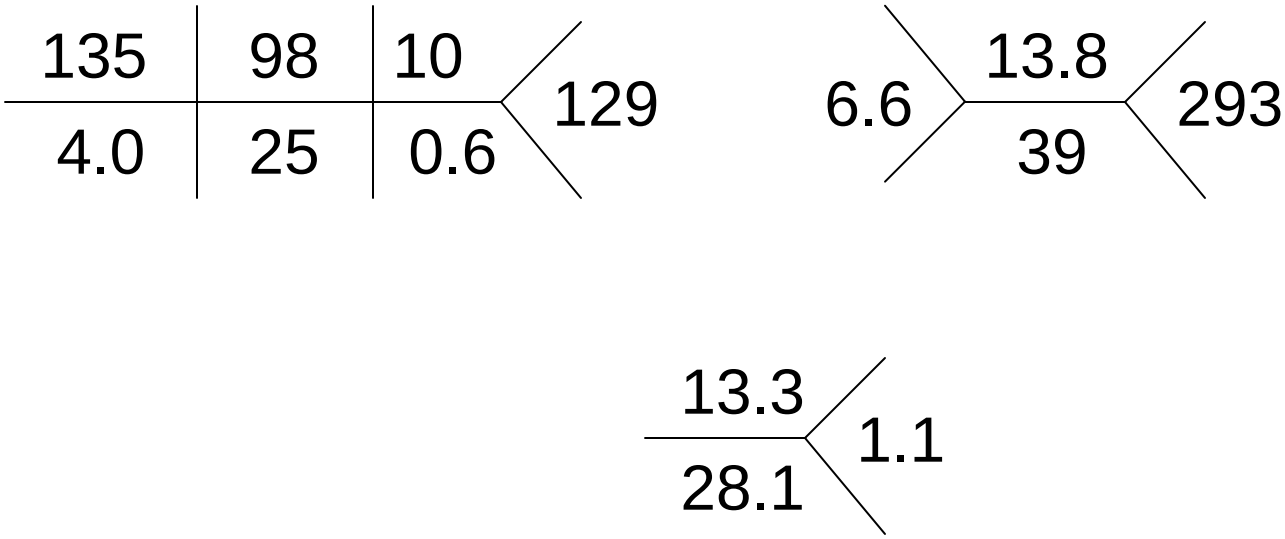
History

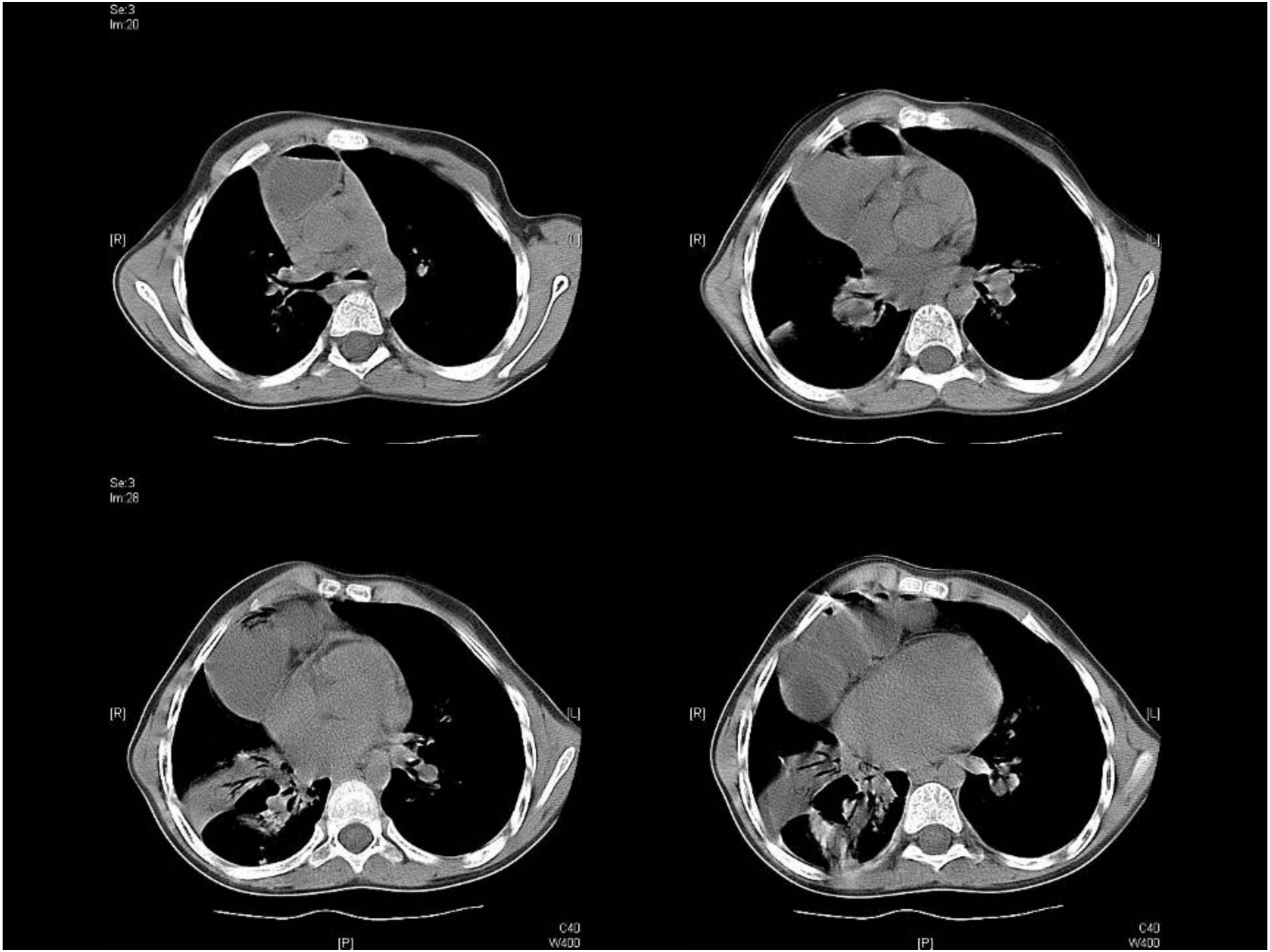
- 8 y.o. boy admitted on 2/17/06 with a diagnosis of severe gastroesophageal reflux disease. He was scheduled to undergo an elective revision of his colonic interposition graft performed 7 years earlier for esophageal atresia. He had multiple prior admissions for aspiration pneumonia and received tube feedings for failure to thrive.
- PMH: Asthma, GERD
- Allergies: latex
- Birth Hx: 32 ½ weeks, polyhydramnios
- PSH: esophagostomy and gastrostomy ’97, colonic interposition ‘98
- Medications: Advair, Singular, Prevacid, Albuterol

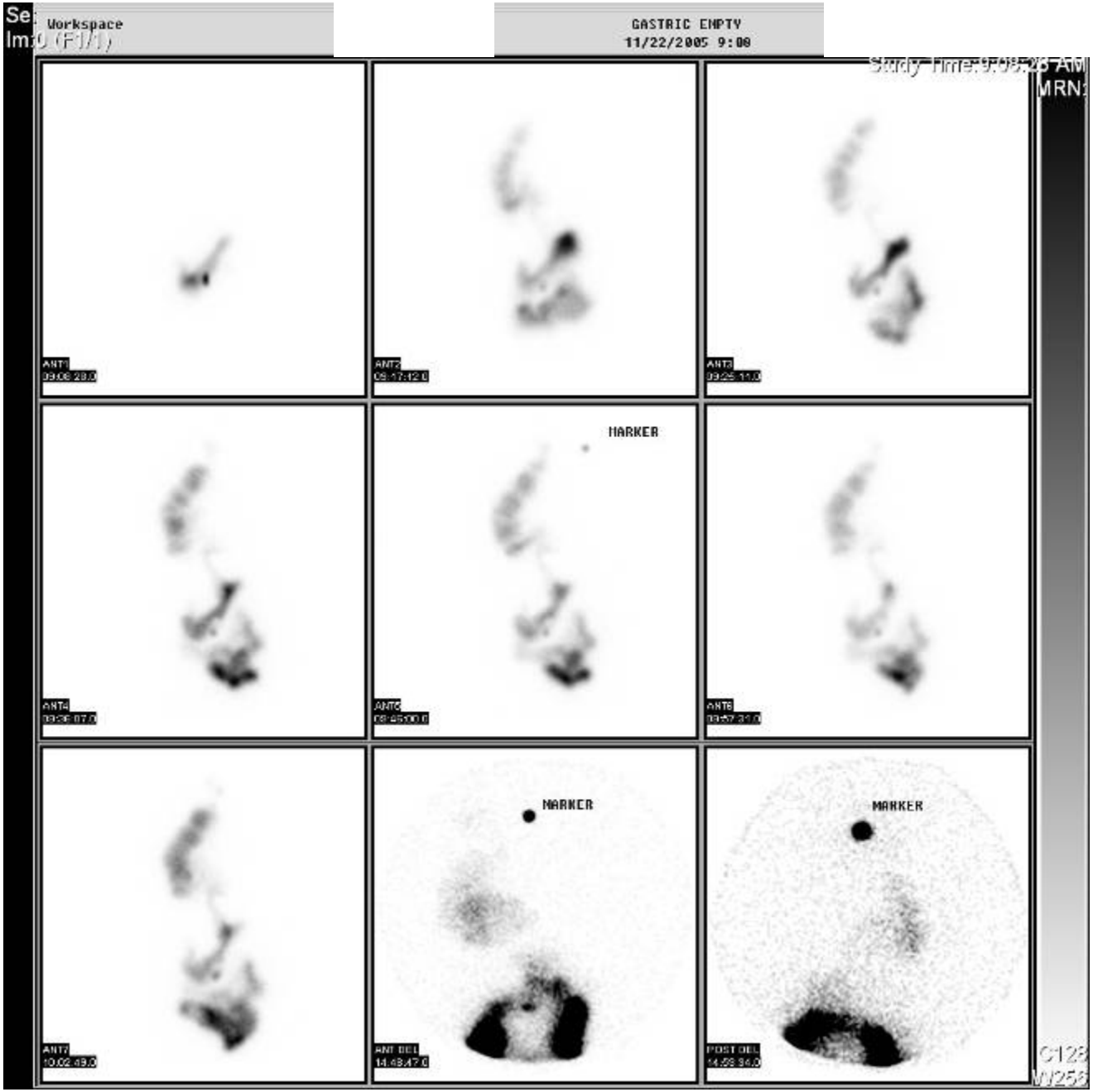
Physical

- Wt 64lbs 75%
- Cardiac: no murmurs
- Chest: CTA b/l
- Abd: well healed upper midline scar, RUQ feeding tube, NT/ND, no masses
- Genitalia: normal, testes descended b/l
- Rectal: normal tone
- Ext: no c/c/e

Laboratory Values

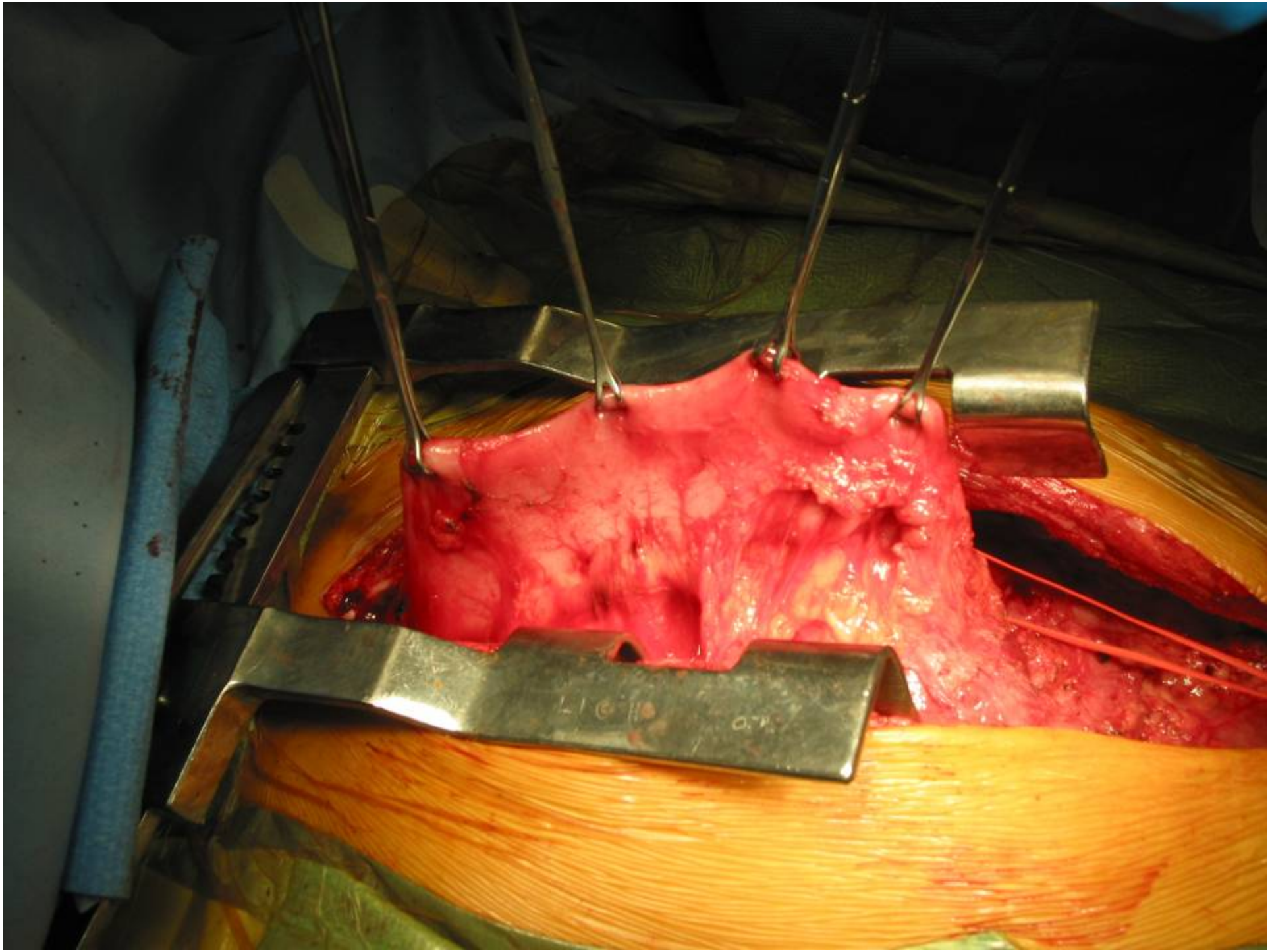


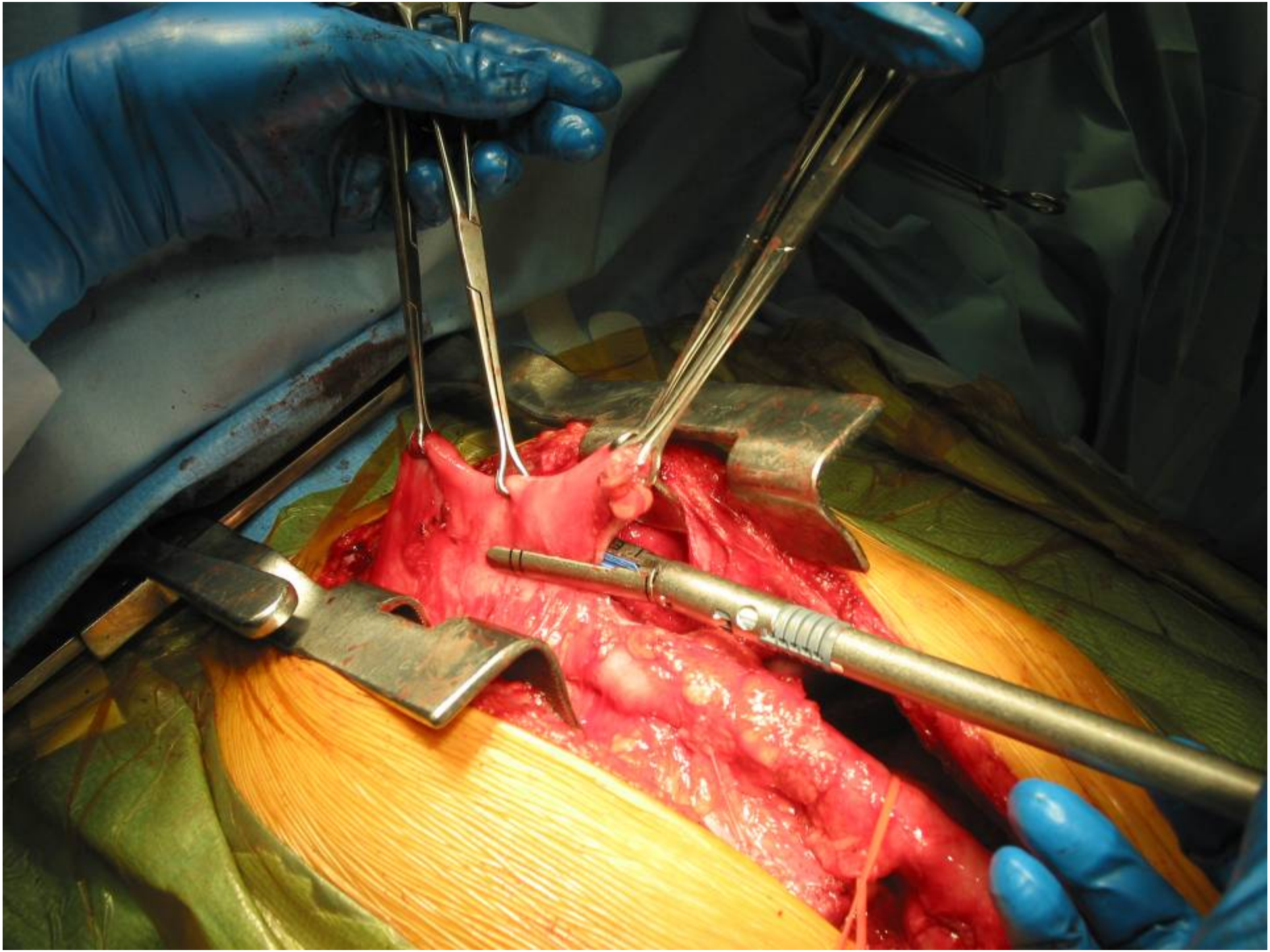

Se:3
Im:20
[R]
[L]
Se:3
Im:28
[R]
[L]
[P]
C40
W400
[R]
[L]
[P]
C40
W400

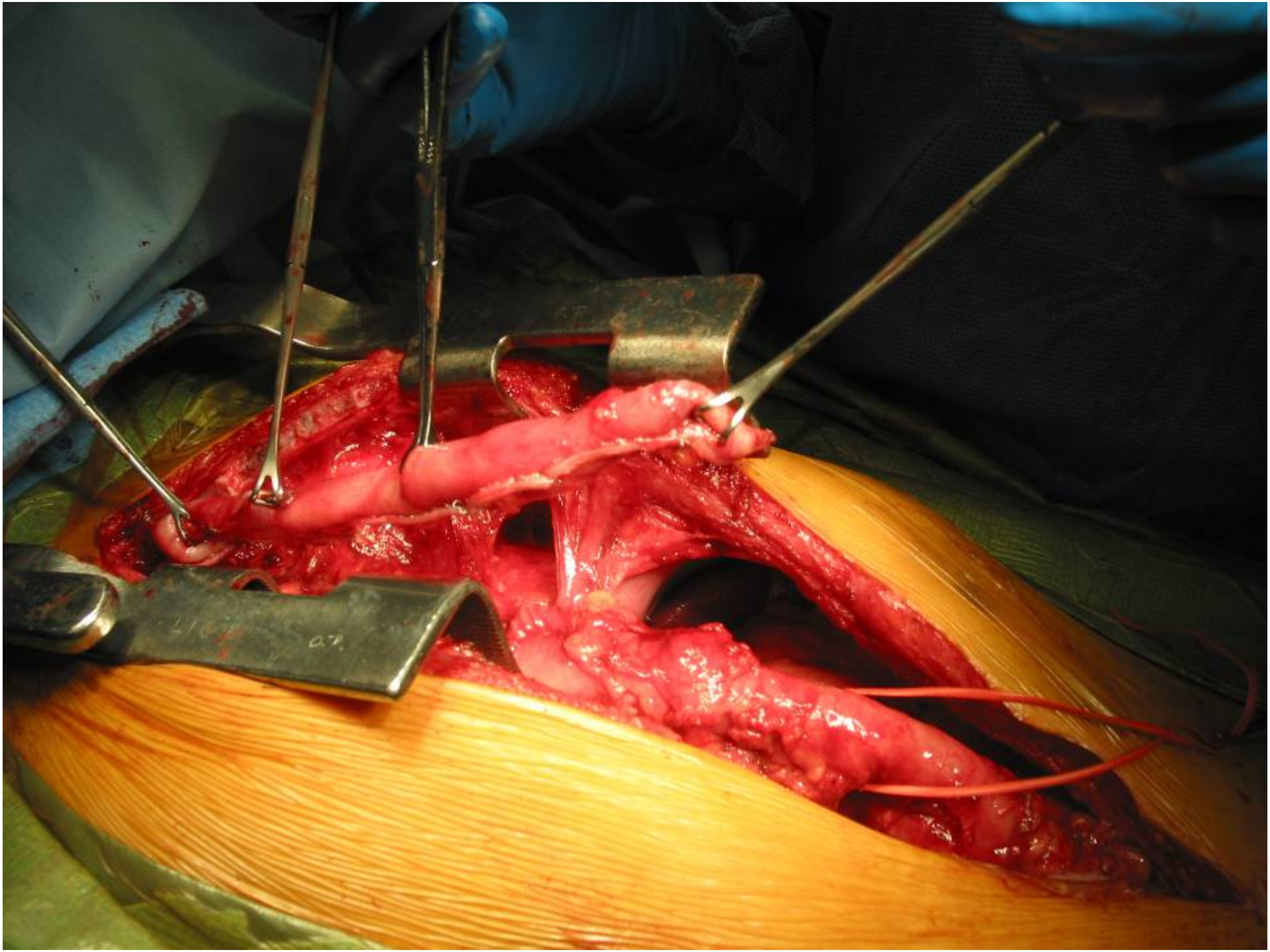

Workspace
GASTRIC EMPTY
11/22/2005 9:08
MARKER
MARKER
MARKER

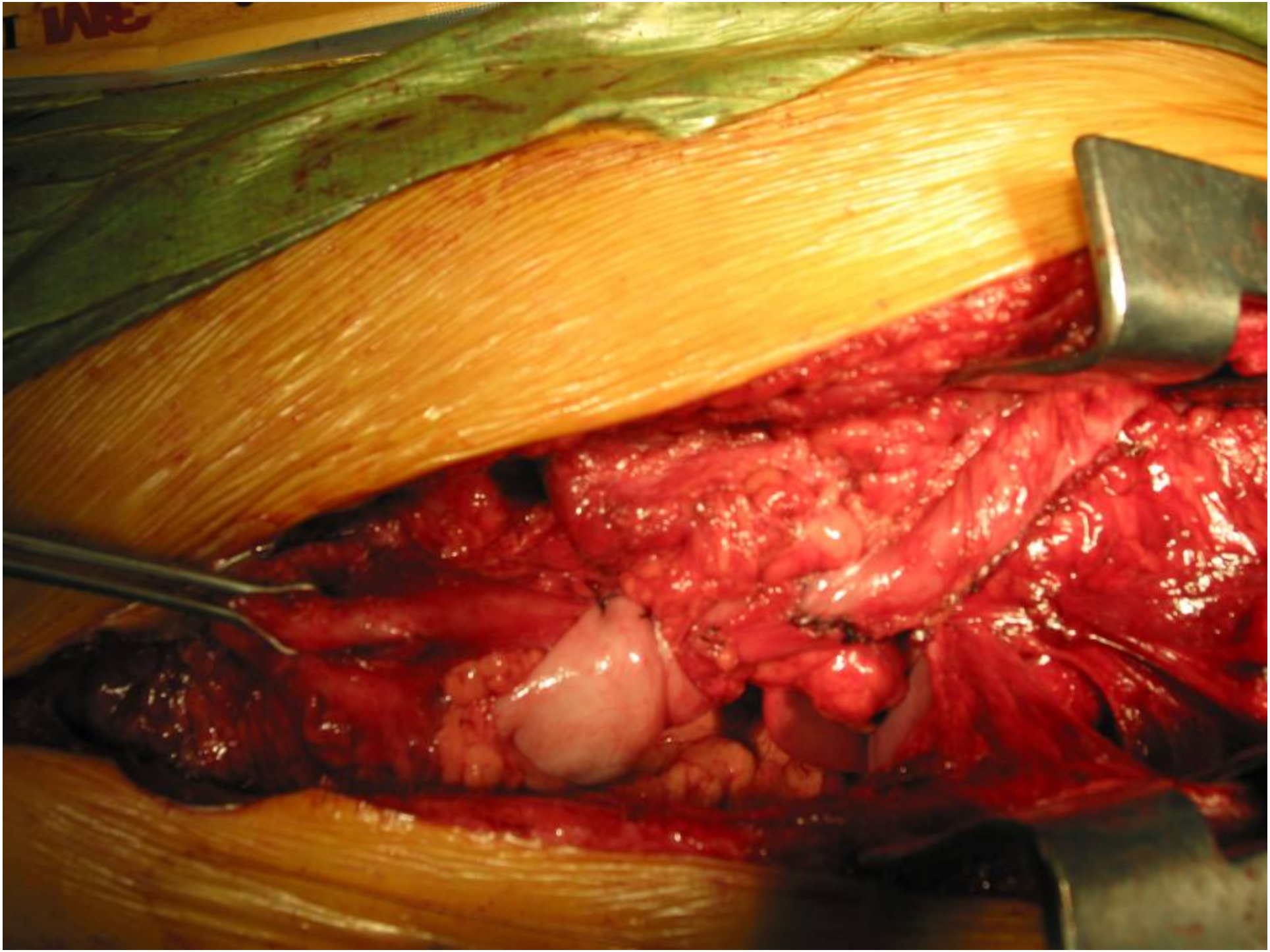
Intraop Events

- Procedure: thoracoabdominal exploration(median sternotomy), tapering coloplasty, partial resection colon interposition, Nissen fundoplication, Stamm gastrostomy
- EBL: 50cc
- Fluids: 1700cc
- OR time: 5 hours



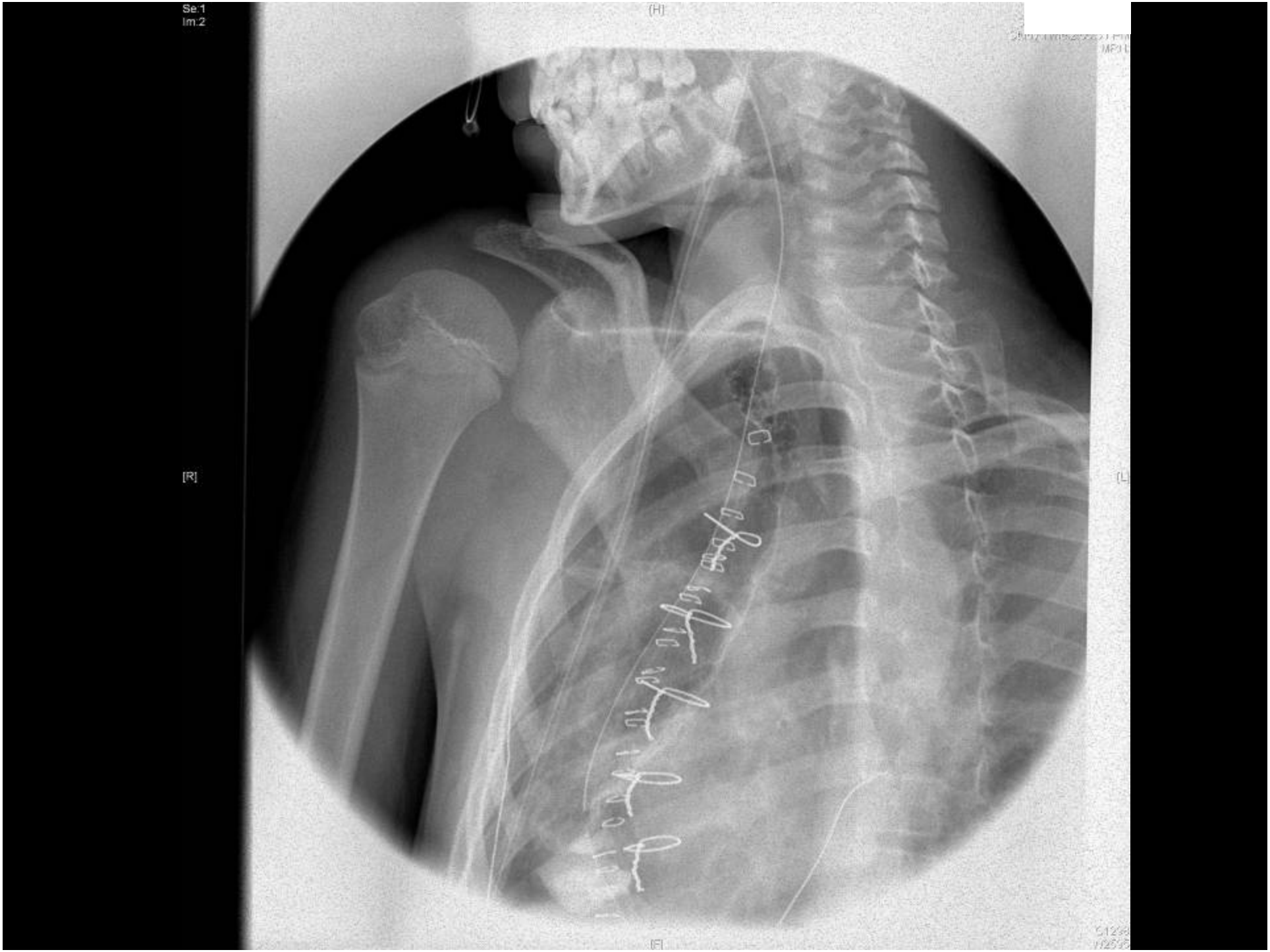






Postop Course

- POD#1 - Extubated in PICU
- POD#6 – Esophogram and return of bowel function
- POD#7 - Tube feeding started
- POD#8 - Full liquids started
- POD#15 – Discharged home tolerating soft diet


Se:1
Im:2
[H]
[R]
[L]
[F]

Management of Esophageal Atresia

Historical Perspective

- 1670 – Durston - First reported case in conjoined twins
- 1888 – Steele - First attempted surgical correction of pure EA by gastrostomy
- 1939 – Levin and Ladd - First survivors of staged repair of EA-TEF

Historical Perspective

- 1941 – First successful primary repair - Haight
  - Left extrapleural approach
  - Primary single layer anastomosis
  - Fistula ligation
- “In recent years there has been no more dramatic advance in surgery than that which has taken place in the treatment of congenital atresia of the esophagus.” Gross R.

Dr. Cameron Haight

Historical Perspective

- Gradual increase in survival rates from 1941 to present 95-99%
- Advances in neonatal intensive care, nutritional support, surgical management, antibiotic therapy

Epidemiology

- Incidence 1 in 3,570 to 4,000
- Slightly higher frequency in males
- 85% cases with tracheoesophageal fistula
- Higher rate low birth weight and preterm birth

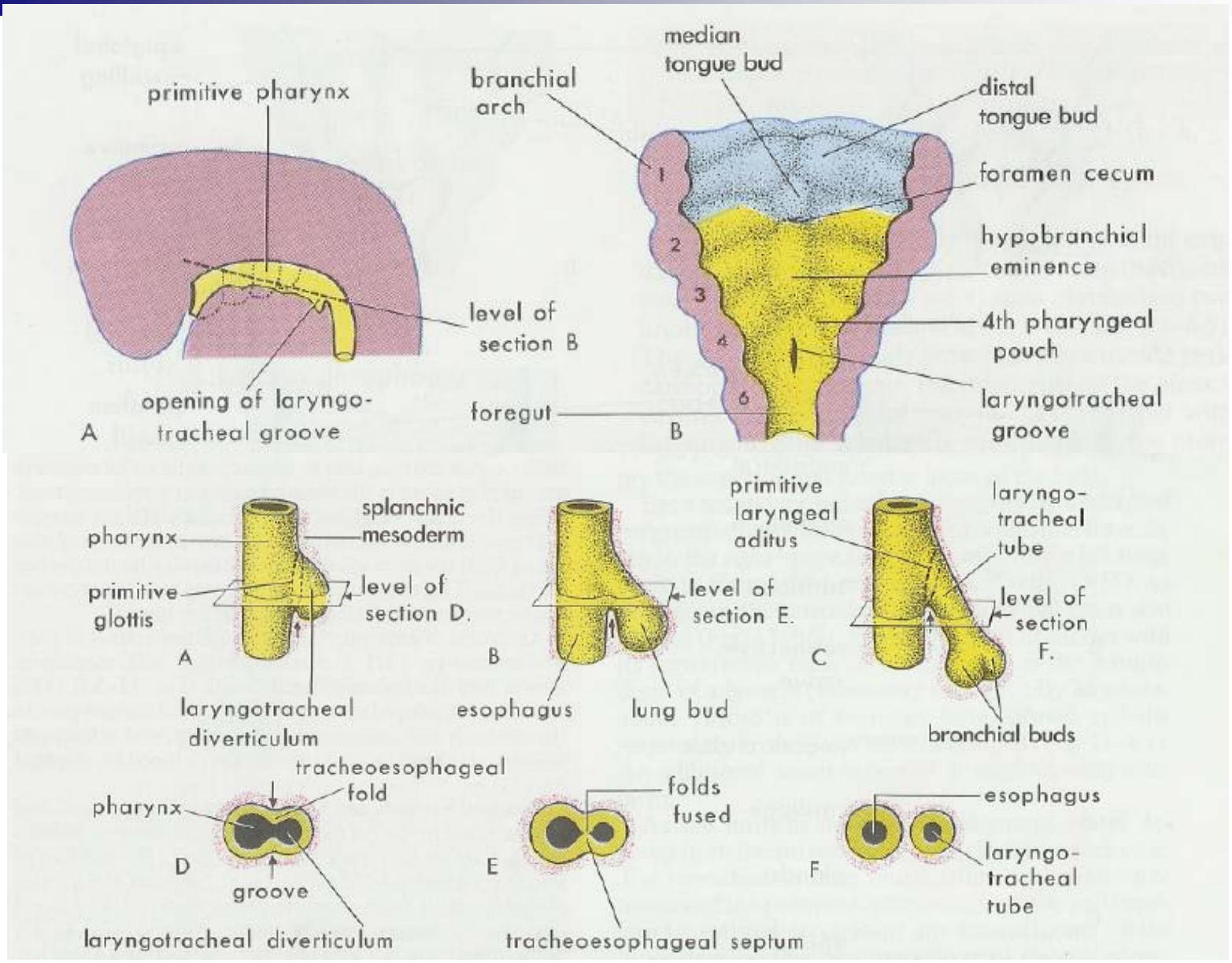
Epidemiology

- 57.3% with atresia only, 36.4% with other major malformations, 6.3% chromosomal anomalies*
- 1 in 10,000 white births 0.55 in 10,000 non white births
- Higher rate in monozygotic twins *

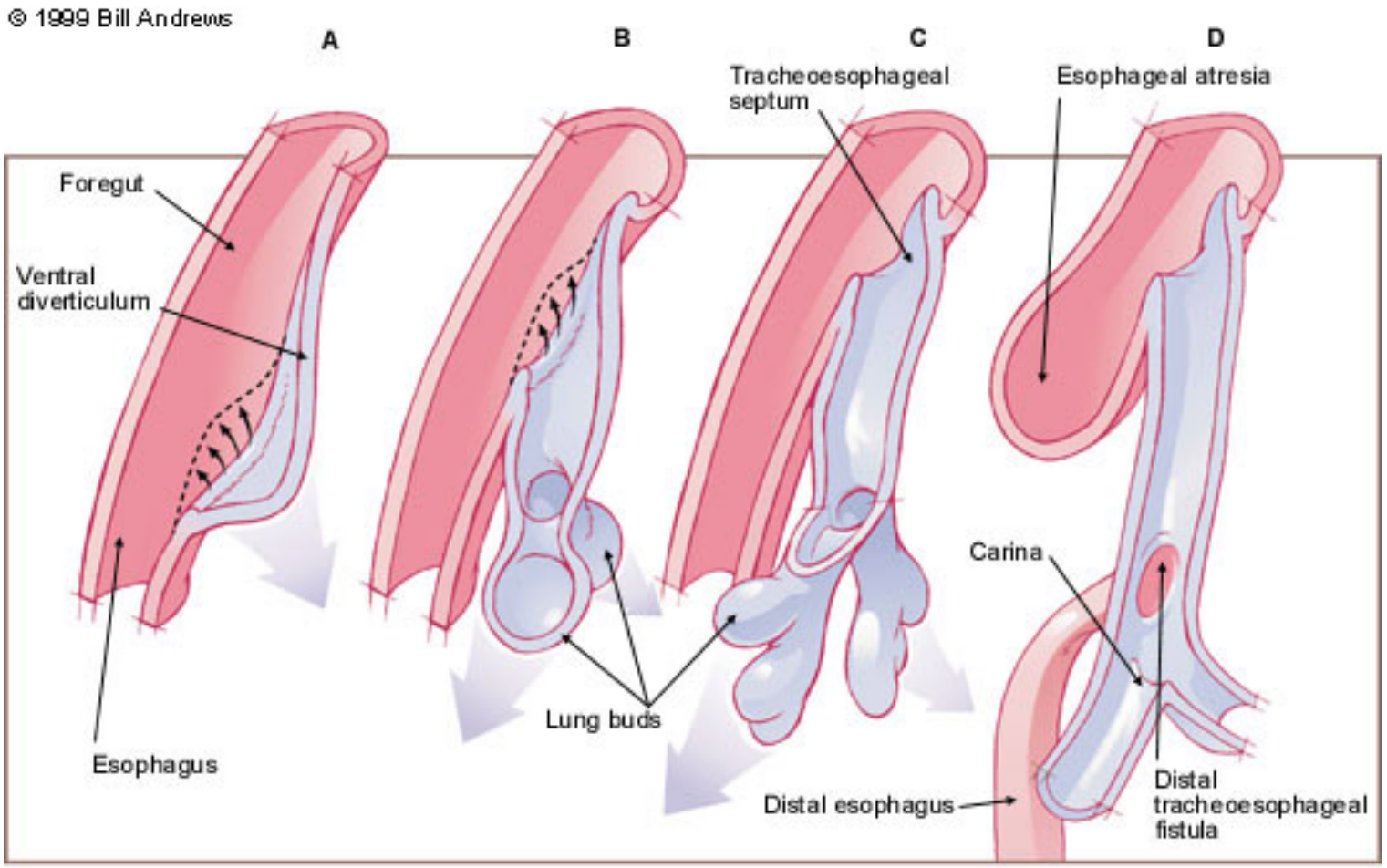
* Robert E, et al. *Reproductive Toxicology*, Vol 7, pp 405-421, 1993

Embryology

- Derived from the primitive foregut
- 4th week of gestation tracheosophageal diverticulum forms from the laryngotracheal groove
- Tracheoesophageal septum develops during the 4th to 5th weeks
- Elongates with descent of heart and lungs
- 7th week reaches final relative length



Moore KL: The Respiratory System. *The Developing Human Being*. 5th ed. Philadelphia, WB Saunders, 1993



Clark DC. *American Family Physician*, 59(4), 1999

Etiology

- Aberration at 26-32 days gestation
  - Differential growth rate
  - Cellular differentiation
  - Apoptosis
- Notochord abnormalities – sonic hedgehog-Gli signaling pathway
- Neural crest abnormalities

Vleesch Dubois VN, et al., *Eur J Pediatr Surg* 2002;12(2):83-89

Presentation/Diagnosis

- Prenatal ultrasound
  - Polyhydramnios (1 in 12)
  - Small or absent stomach
  - Distended blind esophageal pouch
- Prenatal MRI
  - Blind esophageal pouch

Presentation/Diagnosis

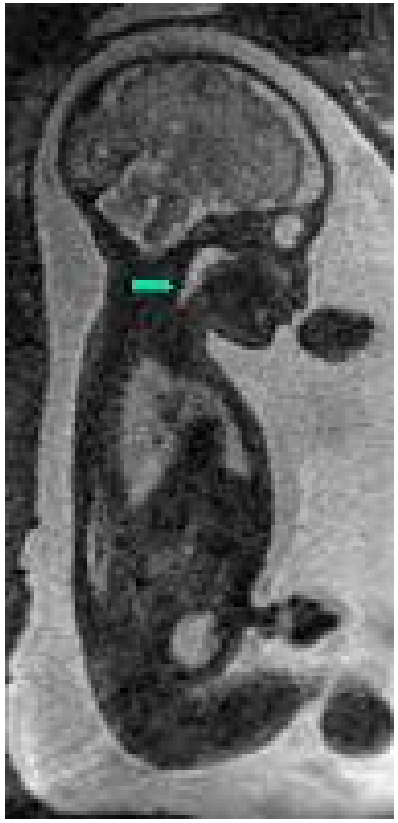
- Postnatally
  - Excessive drooling
  - Fine frothy bubbles in mouth and nose
  - Choking, coughing, cyanotic episodes, and regurgitation with feeding
  - Inability to pass 10Fr OGT

Imaging

- Prenatal ultrasound
  - Good screening test
  - 42% sensitive
  - Polyhydramnios in combination with small or absent stomach
  - No significant role postnatally

Imaging

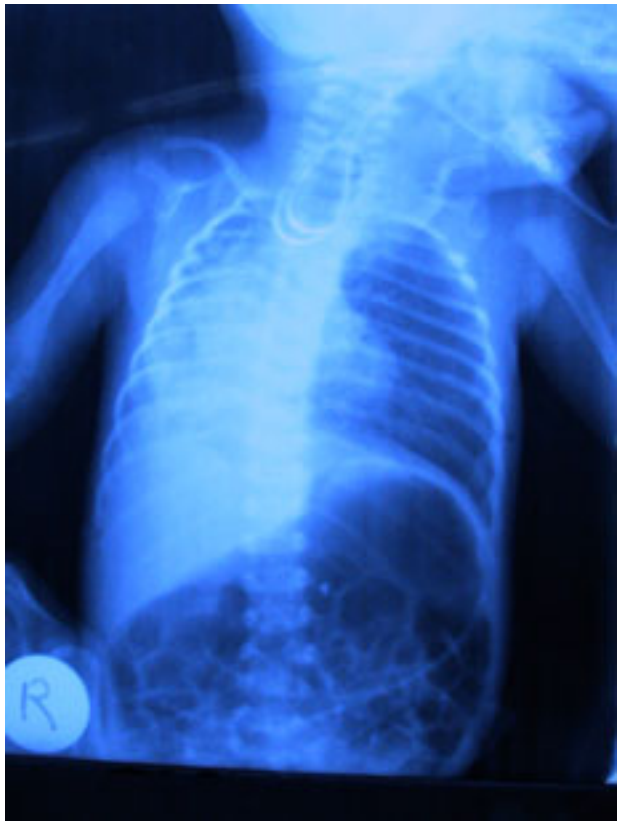
- Prenatal MRI
  - Following positive US findings
  - 100% sensitivity and 80% specific
  - Positive if esophagus absent in mid chest
  - No significant role postnatally



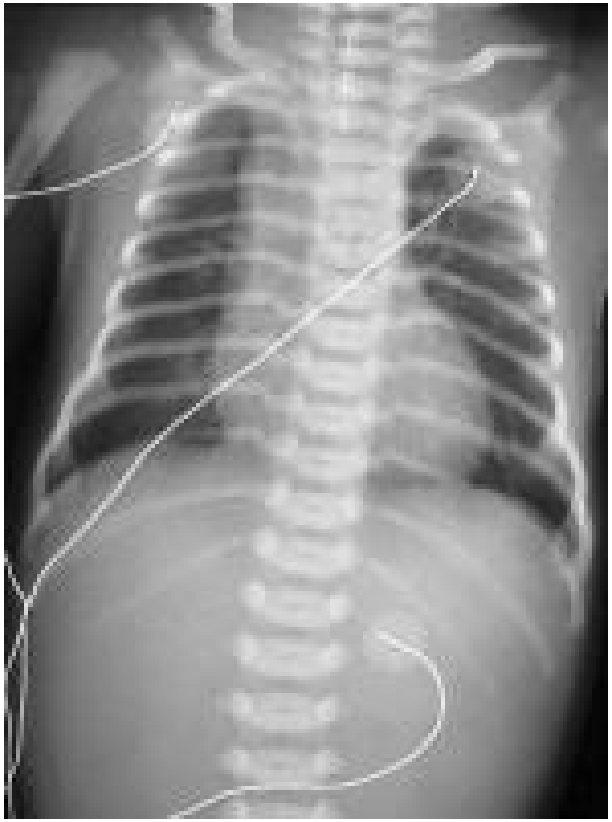
Imaging

- Plain Radiography
  - Confirmatory test
  - OGT in esophageal pouch
  - Presence or absence of gas in the abdomen
  - Assess gap length
  - Anomalies
    - Pneumonitis
    - Atelectasis
    - Cardiac
    - Vertebral
    - Aortic arch

Imaging



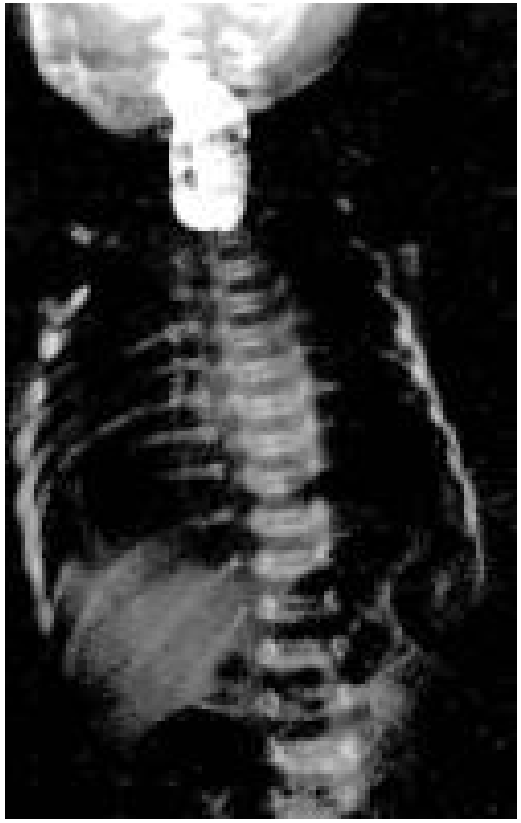
Coiled OGT



Gasless Abdomen

Imaging

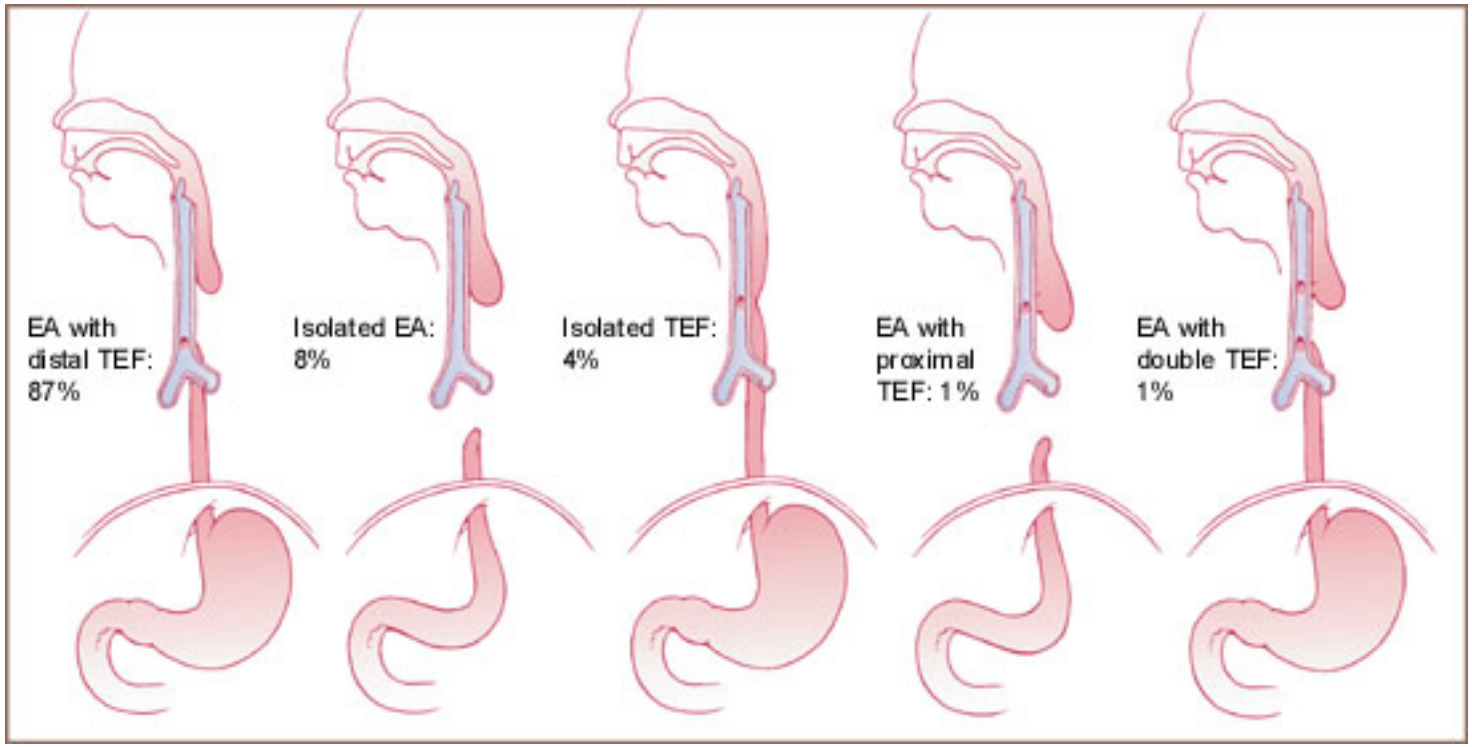
- Contrast studies
  - Rarely needed
  - Identification of proximal fistula
  - Measuring gap length
  - Should use water soluble contrast
  - Perform under fluoroscopy



Imaging/Diagnostics

- Bronchoscopy
- Esophagoscopy
- CT
- MRI

Classification



Clark DC. *American Family Physician*, 59(4), 1999

Associated Anomalies

- VACTERL – vertebral, anorectal, tracheoesophageal, radial, renal
- Trisomy 18 and 21 (7%)
- Congenital heart disease (25%)
- Urinary tract (22%)
- Orthopedic – vertebral and radial (15%)
- GI – i.e. duodenal atresia, imperforate anus (22%)

Management

- Minimal handling to minimize gastric distention and regurgitation
- NPO!!
- Avoid bag-mask ventilation
- Maintain in partial upright position 45°
- Repeated upper esophageal pouch suctioning minimum q10min or low continuous
- Transfer to tertiary pediatric institution for management and definitive care

Management

- IV Tx - 10% dextrose with hypotonic NaCl solution
- Vitamin K analog preoperatively
- Broad spectrum antibiotics
- TPN if surgery delayed for several days

Preoperative Investigations

- Plain radiograph of torso ± contrast
- Echocardiography
- Renal ultrasonography
- Bronchoscopy – selectively
- Esophagoscopy - selectively

Management of Anomalies

- Congenital heart disease
  - Non-duct-dependent conditions – early EA repair
  - Duct dependent disease – PGE1 infusion – EA repair when stable followed by cardiac surgery
  - Right aortic arch – EA through left thoracotomy

Management of Anomalies

- Urinary Tract
  - Reflux associated nephropathy
  - R/O bilateral renal agenesis or severely multicystic dysplastic kidneys
  - Renal scan if absent on US

Management of Anomalies

- Gastrointestinal
  - Careful anorectal exam – EA repair followed by anorectal anomaly
  - Duodenal atresia – combined EA repair duodenoduodenostomy
  - High imperforate anus – combined EA repair colostomy

Management of Anomalies

- Chromosomal
  - EA repair contraindicated in trisomy 18
  - 70% mortality with EA + major chromosomal anomaly
- Orthopedic – no contraindication to EA repair
  - Vertebral i.e. Hemivertebrae – progressive scoliosis
  - Radial club hand
  - Absent thumb
  - Hip dislocation
  - Talipes equinovarus

Surgical Therapy

- Determinants
  - Type of esophageal anomaly
  - Condition of infant
  - Other congenital anomalies present

Surgical Therapy

- Staged Approach
  - Pure EA and select EA and TEF
  - Stamm gastrostomy
  - Delayed fistula division and esophageal reconstruction

Surgical Therapy

- EA and TEF
  - Fistula division with primary esophageal anastomosis
  - Right thoracotomy via 4th ICS
  - Fistula divided close to trachea with air-tight ligation
  - Mobilization of proximal segment with circular myotomy if extra length required
  - Single layer closure with absorbable suture knots internal
  - Feeding tube placed across anastomosis

Surgical Therapy

- Pure esophageal atresia
  - Delayed repair gastrostomy then primary anastomosis
  - Esophageal lengthening
    - Bougienage
    - Circumferential or spiral myotomy
    - Proximal and distal esophageal mobilization
    - Multistaged extrathoracic lengthening
  - Cervical esophagostomy
    - Failed repair
    - Primary anastomosis impossible

Surgical Therapy

- Esophageal replacement
  - Colon –
    - Right, left, or transverse
    - Retrosternal or posterior mediastinal
    - Passive conduit

Colon Interposition

- Pros
  - Acts as a conduit antiperistaltically or isoperistaltically
  - Good vascular supply via marginal artery
  - Can be placed in esophageal bed of posterior mediastinum
  - Has mucous shield, which protects against reflux
  - Responds to acid with a peristaltic rush for clearance
  - Minimizes/eliminates tension on the upper and lower esophageal segments

Minkes RK, Congenital Anomalies of the esophagus. *Emedicine*, May 2006

Colon Interposition

- Cons
  - Requires 3 anastomoses
  - Empties more slowly than the esophagus
  - Requires preoperative bowel preparation
  - Long surgical procedure with extensive mobilization
  - Dilates and becomes redundant over time
  - Slows food transit

Minkes RK, Congenital Anomalies of the esophagus. *Emedicine*, May 2006

Gastric Tube

- Reversed (antiperistaltic) tube
  - More common
  - Supplied by left gastroepiploic artery
- Nonreversed (isoperistaltic) tube
  - Supplied by the right gastroepiploic artery

Minkes RK, Congenital Anomalies of the esophagus. *Emedicine*, May 2006

Gastric Tube

- Pros
  - Does not become dilated, tortuous, or redundant
  - Less risk of ischemia because of robust blood supply
  - Simplified construction through stapler use
  - Has a favorable anatomic location in the upper abdomen
  - Requires fewer anastomoses
  - Has comparable diameter and occupies less space in the thorax and neck
  - Has only one suture line
  - Requires no bowel preparation and is a faster procedure

Minkes RK, Congenital Anomalies of the esophagus. *Emedicine*, May 2006

Gastric Tube

- Cons
  - Extensive gastroesophageal reflux
  - Leaves a small gastric reservoir
  - Creates a long suture line
  - May result in gastric outlet obstruction
  - May be unable to reach high in neck
  - Difficult to place in posterior mediastinum

Minkes RK, Congenital Anomalies of the esophagus. *Emedicine*, May 2006

Gastric Transposition

- Pros
  - Readily available and easily mobilized stomach
  - Involves single anastomosis
  - Adequate length available
  - Has excellent blood supply
  - Involves a technically easy procedure
  - Has low incidence of leaks and strictures

Minkes RK, Congenital Anomalies of the esophagus. *Emedicine*, May 2006

Gastric Transposition

- Cons
  - Large bulky possibly causing space problems intrathoracically
  - Reflux
  - Possible stricture or aspiration due to lack of gastroesophageal valve
  - Poor gastric emptying
  - May affect pulmonary function
  - Results in depleted iron stores causing anemia
  - May not reach as high in neck as other methods because of blood supply

Minkes RK, Congenital Anomalies of the esophagus. *Emedicine*, May 2006

Jejunum

- Pros
  - Jejunal caliber similar to that of normal esophagus
  - Functions as reliable food transporter
  - Results in low incidence of leaks and strictures
  - Functions as an effective gastroesophageal barrier
  - Does no require a bowel preparation

Minkes RK, Congenital Anomalies of the esophagus. *Emedicine*, May 2006

Jejunum

- Cons
  - Length of conduit limited by blood supply
  - Infarction commonly resulting from passage through chest
  - Procedure more technically difficult
  - Requires 3 anastomoses
  - Has high peptic ulcer susceptibility
  - Blood supply lacking marginal artery
  - Has high failure rate

Minkes RK, Congenital Anomalies of the esophagus. *Emedicine*, May 2006

Complications

- Anastomotic leakage 14-21%
- Fistula recurrence 3-14%
- Esophageal strictures 40%
- Gastroesophageal reflux 40-70%
- Tracheomalacia 10-20%
- Esophageal dysmotility

Minkes RK, Congenital Anomalies of the esophagus. *Emedicine*, May 2006

Thoracoscopic Repair of Esophageal Atresia and Tracheoesophageal Fistula

A Multi-Institutional Analysis

George W. Holcomb III, MD, MBA, Steven S. Rothenberg, MD,§ Klaas M. A. Bax, MD,†*
*Marcelo Martinez-Ferro, MD,‡ Craig T. Albanese, MD,‖ Daniel J. Ostlie, MD,**
David C. van Der Zee, MD,† and C. K. Yeung, MD¶

- 104 patients with EA and distal TEF only
- Mean operative time 129.9 min, mean days of hospitalization 18.1
- 5 operations converted to open (4.8%)
- 11.5% developed early leak
- 31.7% required esophageal dilatation
- 24% required required fundoplication later
- 3 deaths

Annals of Surgery 2005;242:422-430

Summary

- Once a death sentence EA and TEF has a survival rate approaching 100% with advances in neonatal care and pediatric surgery
- Primary esophageal repair is the accepted approach if possible
- The selection of esophageal conduits should be made on an individual basis