Pediatric GI Disease in Adults
SGNA Spring Conference
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Clinical Associate Professor of Pediatrics
Pediatric GI Disease in Adults

- Esophageal Atresia
- Duodenal Atresia
- Hirschsprung’s Disease
- Biliary Atresia
- Post Fontan Hepatopathy
Criteria for Review

- Condition limited to pediatric patients
- Successful intervention possible
- Primary adult complications involve GI tract
- Long term outcome data available
Esophageal Atresia

- 1:2500-1:4500 live births
- Possible abnormality of HOXD genes
- Associated anomalies
  - VACTERL
  - Other intestinal anomalies
  - CHARGE syndrome
  - Trisomy 21
  - McKusick-Kaufman syndrome (urogenital/polydac)
  - Fanconi syndrome
Esophageal Atresia

- Polyhydramnios
- Vomits first feed
- Failure to pass ng tube
- Abdomen usually distended
Esophageal atresia
Esophageal Atresia

- EA with distal TEF: 87%
- Isolated EA: 8%
- Isolated TEF: 4%
- EA with proximal TEF: 1%
- EA with double TEF: 1%
Esophageal Atresia

- 1894 Bircher-Skin tube on ant. thorax
- 1907 Roux- Jejunum tunneled subcut.
- 1920’s Subcutaneous colon swing
- 1943 Haight-Primary intrathoracic repair
- 1948 Intrathoracic colon interposition
- Routine intrathoracic repair of EAT began in which era?
Mid 1960’s
## Esophageal Atresia - Survival

<table>
<thead>
<tr>
<th>Criteria</th>
<th>Survival</th>
</tr>
</thead>
<tbody>
<tr>
<td>BW &gt; 1500gm</td>
<td>283/293 (97%)</td>
</tr>
<tr>
<td>No cardiac defect</td>
<td></td>
</tr>
<tr>
<td>BW &lt; 1500gm or major cardiac defect</td>
<td>41/70 (59%)</td>
</tr>
<tr>
<td>BW &lt; 1500gm and major cardiac defect</td>
<td>2/9 (22%)</td>
</tr>
</tbody>
</table>

Esophageal atresia

- Squamous cell carcinoma (38yrs)
- Adenocarcinoma (20, 35, 46yrs)
- Gastroesophageal reflux/esophagitis
- Carcinoma in skin tube conduit
- Chest wall deformity
- Barrett esophagus in esophageal remnant
Barrett’s Esophagus

- Intestinal Metaplasia of esophagus
- Results from chronic injury
- Most reports in children with neurodevelopmental disability
- In adults, risk of adenoCa-40-125x
- Scattered reports of adenocarcinoma in children-Associated with Barretts
- ?Reverses with fundoplication
69 patients operated 1972-1990
7 had colon interposition
Mean follow-up 10.5 yrs
42 children >10 yrs
  40% dysphagia
  31% GERD
  17% wt < 5th %ile
  10% Recurrent resp infections
  7% choking spells
Experiences of a Parent Support Group with the Long-term Consequences of Esophageal Atresia-Schier J Ped Surg 2001

- 128 patients
- Follow-up 10 to 34 years (median 14 yrs)
- Recurrent lung infections 80%
- GE reflux 46%
- ‘Below normal weight’ 30%
- Dysphagia 30%
- Barretts esophagus 7%
- ‘Unorthodox food combinations’ 13%
Barrett’s Esophagus in Gastroesophageal Reflux and Adults Born with Esophageal Atresia-Krug Am J Gastro 1999

- 69 patients with primary anastomosis 1971-78
- 24 died (5-aspiration)
- 39 traceable survivors
  - 30/39 dysphagia
  - 13/39 GE reflux
  - 2/34 Barrett’s esophagus
Esophageal Disease in National Cohort

- 502 cases born 1949-1978 in Finland
- All cases tracked through cancer registry 1967-2004
- 230 patients died before 1967
- 272 remaining patients-No Esophageal cancers
- 101 re-studied-34% GER, 85% dysphagia, 11% Barretts, 8% stricture
- Manometry-Non propogating swallows
Some 60’s Rockers tolerate the ravages of time…
Others have not!
Duodenal Atresia

- 1:5000-1:10000 live births
- 30% associated with Trisomy 21
- Cong heart defects without T21 25%
- Detected antenatally in 32-57%
- Present with bilious emesis in most cases
- “Double bubble” sign
Double Bubble

- The very first bubble gum was invented by Frank Henry Fleer in 1906. He called it Blibber-Blubber.

- In 1928, an employee of the Frank H. Fleer Company, Walter Diemer invented the successful pink colored Double Bubble, bubble gum.
Double Bubble
Duodenal Atresia Repair

- First reported repair 1905-Vidal
- First DJ repair 1914-Ernest
- Duodenoduodenostomy 1973-75- Grosfeld, Kimura
Duodenal Atresia and Stenosis: Long term follow-up over 30 years - Escobar J Ped Surg 1004

- 169 pts 1972-2001
- Retrospective with average follow-up 6 yrs
- 27 has at least one other GI surgery over next 6 years (13 fundos)
- 10 Deaths (5 from complex cardiac malformations)
Duodenal Atresia-Late follow-up
Kokkonen J Ped Surg 1988

- 41 pts chosen randomly 15-35 years (mean 22 yrs) after surgery for further evaluation
- 13 reported persistent symptoms
- Upper GI normal in 2, massively dilated duodenum in 9
- Endoscopy-Abnormal duodenum in 19
January 1, 2006: The first baby boomers turn 60...

Far out! The old uniform still fits!
Hirschsprung Disease

- Failure of craniocaudal migration of ganglion cell precursors
- Occurs between 5 and 12 weeks of gestation.
- Timing determines length of aganglionic segment.
- Reduced nitric oxide synthase.
Hirschsprung Disease

- 1888 Hirschsprung - Entire colon defect
- 1898 Treves - Sigmoid colostomy
- 1937 Adson - Neuro cause recognized
- 1943 Whitehouse - Absence of ganglion
- 1948 Swenson - Endorectal pullthrough
- 1960 Duhamel - Modification of Swenson
- 1964 Soave - Modification of Duhamel
Adult Diagnosis of Hirschsprung’s

- 20% of patients undiagnosed by age 5 years.
- Life long constipation
- Prominent abdominal distension
- Male predominance
- Barium enema suggests diagnosis
- Complication rate appears higher
  - Stricture
  - Impotence
  - Incontinence
1964 Civil Rights Act

- To enforce the constitutional right to vote.
- To confer jurisdiction upon the federal courts to provide relief against discrimination in public accommodations.
- To authorize the Attorney General to institute suits to protect constitutional rights in public facilities and public education.
- To prevent discrimination in federally assisted programs
- To establish a Commission on Equal Employment
## Hirschsprung Follow-up

<table>
<thead>
<tr>
<th>Follow-up yrs</th>
<th>No.</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt;6 yrs</td>
<td>337</td>
<td>41.4</td>
</tr>
<tr>
<td>6-10</td>
<td>186</td>
<td>22.9</td>
</tr>
<tr>
<td>11-15</td>
<td>96</td>
<td>11.8</td>
</tr>
<tr>
<td>16-20</td>
<td>78</td>
<td>9.6</td>
</tr>
<tr>
<td>21-25</td>
<td>53</td>
<td>6.5</td>
</tr>
<tr>
<td>26-30</td>
<td>22</td>
<td>2.7</td>
</tr>
<tr>
<td>31-35</td>
<td>20</td>
<td>3.7</td>
</tr>
<tr>
<td>&gt;35</td>
<td>12</td>
<td>1.5</td>
</tr>
</tbody>
</table>

Sherman J Pediatr Surg 1989;24:833
## Hirschsprung Follow-up

<table>
<thead>
<tr>
<th>Length of follow-up</th>
<th>&gt;15 yrs</th>
<th>&gt;20 yrs</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal bowel habits</td>
<td>191/207 (92.3%)</td>
<td>118/126 (93.7%)</td>
</tr>
<tr>
<td>No soiling</td>
<td>196/203 (96.6%)</td>
<td>122/124 (98.4%)</td>
</tr>
<tr>
<td>1-3 BM’s/d</td>
<td>184/195 (94.4%)</td>
<td>117/121 (96.7%)</td>
</tr>
</tbody>
</table>

Sherman J Pediatr Surg 1989
## Hirschsprung Follow-up

<table>
<thead>
<tr>
<th>Continence score</th>
<th>No. (%)</th>
</tr>
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<tbody>
<tr>
<td>Normal (score 14)</td>
<td>14 (26%)</td>
</tr>
<tr>
<td>Good (score 10-13)</td>
<td>34 (63%)</td>
</tr>
<tr>
<td>Fair (5-9)</td>
<td>6 (11%)</td>
</tr>
</tbody>
</table>
Hirschsprung Follow-up

Heikkinen J Pediatr Surg 1997;32:1443
Long term Follow-up of Hirschsprung’s

- 183 pts 1963-1989 - Single center - Japan
- 153 survivors surveyed
- 85.7% “satisfactory bowel function”
- 19% soiling
- Ieiri et al J Ped Surg 2010
The Joy of Tech™

by Nitrozac & Snaggy

HELP! SHE’S FALLEN, AND CAN’T GET UP!
SOMEONE CALL 911!
I WILL! I HAVE MY iPHONE WITH ME!

...AND THEN IF YOU TURN IT SIDEWAYS, THE MOVIE GOES WIDESCREEN!

.nd the other iPhone lawsuit.

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joyoftech.com
Extrahepatic Biliary Atresia

- 1:8000-1:15000 live births
- Cardinal features - Jaundice, acholic stools and hepatomegaly
- Usually term and healthy appearing
- 10% have other anomalies (polysplenia)
- Kasai portenterostomy 1959
Kasai Surgery
Extrahepatic Biliary Atresia

- 48 pts (10-33yrs)-All had abnl LFT’s, 6 jaundiced, 11 Varices- (Ohi J Ped Surg 1990)
- 104 consecutive pts tracked after 10 yrs
  - 98 Kasai procedures, 63 deaths, 35 liver tx
  - 19 alive w/o transplant
- 11/122 children alive w/o transplant- (Laurent Gatro 1990)
Long Term EHBA

- 141 Children, 1976-2000, single center, Taiwan-s/p Kasai (Hung JPGN 2006)
  - 10 yr survival rate with native liver 31%
  - Overall 10 yr survival 40.2%

- 80 Patients, 1970-86, Japan (Shinkai JPGN 2009)
  - 10 yr native liver 54%
  - 20 yrs 50% cirrhosis, GI bleeding 17%
Liver Transplantation

- 1963 First liver transplant-3 yr EHBA
- Before 1970 mortality >50% 1 yr
- Cyclosporine 1978
- Prograf-1989
- 2002 500 peds liver transplants/100ctrs
- 5 yr survival 80-90%
1981
Tricuspid Atresia

- BT shunt, Glenn shunt
- Fontan-1974
  - Anastamosis of right atrium to pulmonary artery.
  - IVC blood flow separated from heart
  - GI complications from increased IVC pressure-GI protein loss and liver disease
Fontan Procedure

- Late decline in survival-334 pt retrospective (Fontan-Circulation 1990)
- Autopsy series-7/9 pts had abnl liver histology, 4 cirrhosis (Ghaferi, J Thor CV Surg 2005)
- Retrospective 12 pts (15-43 yrs)-7/12 cirrhosis, 4/12 varices (Kieswetter Heart 2007)
- 3 case reports of hepatocellular Ca
1955 No Hair
2010 No Hair
"...Transition is a process, not an event. The actual process should be gradual, occurring in harmony with adolescent and family development. While there is not one current model, whenever it occurs, communication among pediatric and adult providers, parents and youth is critical."

--From the National Center for Youth with Disabilities
Transition of Health Care

- **3 Competencies** (Olson 2004)
  - Developing practices that encourage the patient to accept responsibility for his/her healthcare
  - Planning for the future with families
  - Linking families to share information and experiences
- Educating adult providers
Specific Goals for Transition
AAP (Pediatr 2002;110:1304)

- Develop a life plan
- Create folder/file with complete medical and surgical records.
- Use transition worksheets-dscc.uic.edu
- Provide same primary care services
- Plan for continuous health insurance coverage
You're right... A nap seems like a much better idea...

Scene from "The Tortoise and The Hare II: The Middle Age Years"