Author's response to reviews

Title: Application of a plain abdominal radiograph transition zone (PARTZ) in Hirschsprung's disease: Implications for a single stage transanal pull through

Authors:

Akshay Pratap (akshaypratap2000@gmail.com)
Devendra K Gupta (devendra6@hotmail.com)
Awadhesh Tiwari (atiwari@yahoo.co.uk)
Arvind K Sinha (aksinha@yahoo.com)
Nisha Bhatta (nisha@hotmail.com)
Chandra S Agrawal (drcsagrawal@yahoo.com)
Shailesh Adhikary (sadhikary@hotmail.com)
Anand Kumar (anand_bhu@satyam.net.in)

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Author’s replies

Thank you for your comments on this paper.

I would like to clarify that there is no doubt a rectal biopsy has to be made before proceeding for a pull through, which we have performed in all cases. I also agree with your conclusion that PARTZ is only 92% sensitive leaving a misdiagnosis in 8% of patients. Next, there was no transition zone seen on the barium enema in nearly 33% of the patients in the study. In my mind, if I don't see a contrast enema transition zone (which is especially the case in neonates), I am not really that concerned that the patient may not be an appropriate candidate for a primary pullthrough unless I don't see one even on a plain abdominal radiograph. PARTZ that we see on a radiograph is secondary to the spasm of the aganglionic segment resulting in a cutoff at that level. This is evident even in the absence of thickening of the aganglionic colon wall, a phenomenon responsible for the transition zone to be seen on a contrast enema. The limitations of a contrast enema in newborns have been discussed in detail in this regard, and this is where I believe a simple plain abdominal radiograph is very informative.

Role of laparoscopy or Umbilical incision

I agree with you that once we approach the proximal sigmoid colon we cannot do a transanal pullthrough safely without mobilizing the mesentery and/or mobilizing the splenic flexure. In these patients bowel mobilization either laparoscopically or through an umbilical incision is of immense benefit. In the present study there were 2 patients who were although excluded from the work up showed a long segment involvement. An umbilical incison assisted biopsy was taken from the transition zone and a stoma was subsequently created. I am advocating the use of umbilical incision and have a universal agreement with all of my colleagues. We are very comfortable with this approach, especially with the lack of laparoscopic facilities. This aspect has been incorporated in the manuscript.

There are some surgeons who are of the opinion of performing a laparoscopic biopsy to identify the transition zone pathologically before the start of the dissection. I am not sure that there is a universal agreement with this approach. My own personal opinion is that a laparoscopic or umbilical biopsy should be offered to patients in whom a transition zone either on plain abdominal radiograph or contrast enema is not clearly defined or in those in whom a danger
of potential vascular compromise due undue tension on the pullthrough colon exists. I think with this approach we can be very noninvasive till a point when we are compelled invade the abdomen for ensuring the level of transition zone or facilitate mobilization of colon.

**Issue of Enterocolitis**

Not every baby is a good candidate for a primary pullthrough. However, we do see a lot of children who present with relatively mild enterocolitis. We usually treat them with antibiotics and irrigations, settle it down, and then go ahead and do a primary pullthrough. On the other hand some babies have severe sepsis and poor general condition and are subjected to a diverting colostomy. We adopted Bells criteria of staging enterocolitis, since there is no other scoring system available for Hirschsprung's disease as such. If NEC was suspected staging was performed:

**STAGE I**
- b. Systemic manifestations-temperature instability, lethargy, apnea, bradycardia.
- c. Gastrointestinal manifestations-poor feeding, increasing pregavage residuals, emesis, mild abdominal distension, occult blood.
- d. Abdominal radiographs show distension.

**STAGE II**
- a. Above signs and symptoms plus persistent occult or gross gastrointestinal bleeding; marked abdominal distension.
- b. Abdominal radiographs show significant intestinal distension with ileus; small bowel separation (edema in bowel wall or peritoneal fluid), unchanging or persistent "rigid" bowel loops.

All stage I and Stage II patients were managed conservatively with intravenous fluids, broad spectrum antibiotics, nil oral and rectal washes for a period of 48 to 72 hours. No attempt of a pull through was made even in Stage I disease. If they did not settle after this period a diversion colostomy was done. If they showed signs of improvement we would wait for 10 days before deciding on pull through. Stages III were excluded straightaway because of life threatening sepsis.