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

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Application of a plain abdominal radiograph transition zone (PARTZ) in Hirschsprung's disease: Implications for a single stage transanal pull through
Akshay Pratap, Devendra K Gupta, Awadhesh Tiwari, Arvind K Sinha, Nisha Bhatta, Chandra S Agrawal, Shailesh Adhikary and Anand Kumar

Thank you for consideration of our manuscript for publication in your journal.

We have reviewed the above manuscript according to your reviewer’s comments.

Reviewer # 1 (Dr Jacob Langer)

**Major Compulsory revisions**

1 and 2. The authors have clarified to some extent how the use of the PARTZ helps in the use of a one-stage transanal approach, particularly in cases where the barium enema is unclear. However, the PARTZ is still only 92% accurate, leaving a misdiagnosis of the level of transition zone in 8% of the cases (coincidentally, the same number we reported in reference 6). To my way of thinking, this is still too high, and justifies the use of a preliminary biopsy in every case. The authors have talked about the use of laparoscopy, and correctly pointed out that this technique is not available in many developing countries. However, for precisely this reason, we have advocated the use of an umbilical incision for the biopsy, a technique which is easy and can be done by any pediatric surgeon (Sauer CJE, Langer JC, Wales PW: The versatility of the umbilical incision in the management of Hirschsprung’s disease. Journal of Pediatric Surgery 40:385-89, 2005). This approach should at least be discussed and referenced in this manuscript, since it bears directly on the authors’ conclusions and really nullifies the importance of the PARTZ in determining the level of the transition zone. Why use a 92% accurate X ray finding (that may be dependent on the skill of the radiologist), when there is another option that is 100% accurate and easily performed by any pediatric surgeon?

- 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 incision 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 (see page 9 and 10, reference 14). 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 left with no choice but to invade the abdomen for ensuring the level of transition zone or facilitate mobilization of colon.

### 3. The Bell classification has been used for NEC, not for Hirschsprung's associated enterocolitis. However, the authors are correct that there is no good definition of enterocolitis. I would like to see them specify how the Bell classification was used, ie did they have to be Bell stage 2?

- 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 pregaavage 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.

7. References are better, except for my comments about the umbilical incision above.
   - We have included the comment and the reference in the revised manuscript (see page 10 and reference 17):

**Declaration of competing interests:**
I declare that I have no competing interests.