



(neuronal intestinal dysplasia)

(Hirschsprung)

Hirschsprung

가

가

B

2

1

Hirschsprung

(Fig. 1C).

total colonic agangliosis

3

Hirschsprung

가

가

1

total colonic agangliosis

(Fig. 1D).

B

2

10

가

(ileostomy)

29

가

B

(Fig. 1E).

40

X -

(scout roentgenogram)

가

가

(Fig. 1A).

(Fig.

1B).

1971

Meier - Ruge

가 Hirschsprung

(1).

. 10

Scharli Meier - Ruge

(2).

Fadda

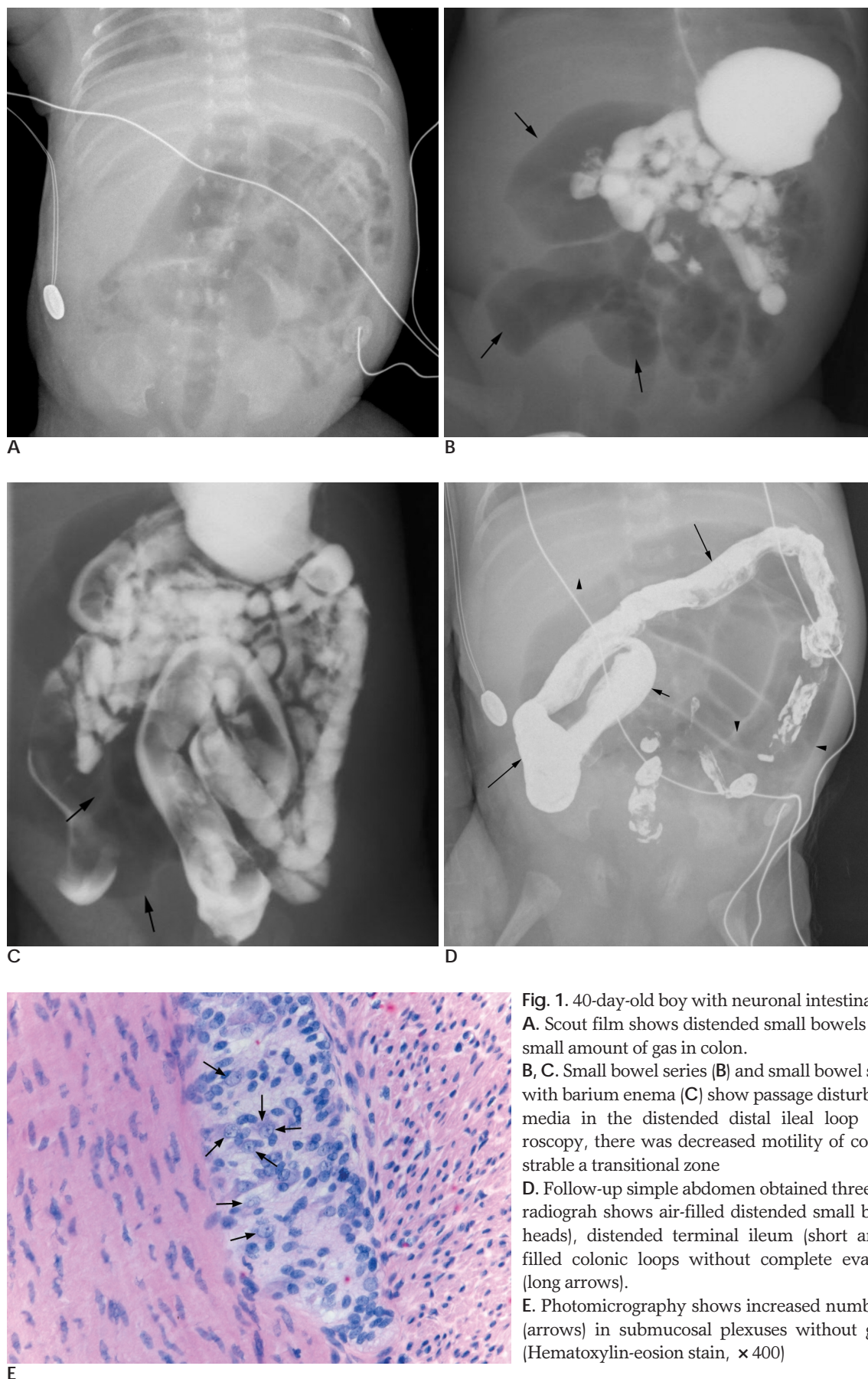
(1)

A B ,

. A

1  
2  
3

2005 11 4 2006 1 10



**Fig. 1.** 40-day-old boy with neuronal intestinal dysplasia  
**A.** Scout film shows distended small bowels filled with gas, and small amount of gas in colon.  
**B, C.** Small bowel series (**B**) and small bowel series combined with barium enema (**C**) show passage disturbance of contrast media in the distended distal ileal loop (arrows). On fluoroscopy, there was decreased motility of colon and no demonstrable a transitional zone  
**D.** Follow-up simple abdomen obtained three days after initial radiograph shows air-filled distended small bowel loops (arrowheads), distended terminal ileum (short arrow) and barium-filled colonic loops without complete evacuation of barium (long arrows).  
**E.** Photomicrography shows increased number of ganglion cells (arrows) in submucosal plexuses without giant ganglion cell. (Hematoxylin-eosin stain,  $\times 400$ )

- 5% . B total colonic agangliosis
- 95% Hirschsprung , Hirschsprung
- A B
- acetylcholinesterase 가 (4).
- A , B 6 6 Hirschsprung (inflammatory bowel disease)
- 가 Hirschsprung (10).
- B Puri (4) 6 , 3 가 B 6 (internal sphincterectomy) Soave (4).
- (3). B 가 0.3 - 40% (4).
- 가 1 , 가
- Motetonico (5)
- Hatanop Boyen (6, 7) Hox11 L1 endotheline (receptor)가
- 가
- Kanamori (8)
- 가
- 1 가 (9) 2
- B 가
- B Hirschsprung (rectosigmoid colon)가
- Hirschsprung (4, 5).
1. Fadda B, Maier W, Meier-Ruge W, Scharli A, Daum R. Neuronal intestinal dysplasia. critical 10-years ' analysis of clinical and biopsy diagnosis. *Z Kinderchir* 1983;38:305-311
  2. Scharli A, Meier-Ruge W. Localized and disseminated forms of neuronal intestinal dysplasia mimicking Hirschsprung 's disease. *J Pediatr Surg* 1981;16:164-170
  3. Schmittenbecher PP, Gluck M, Wiebecke B, Meier-Ruge W. Clinical long-term follow -up results in intestinal neuronal dysplasia (IND). *Eur J Pediatr Surg* 2000;10:17-22
  4. Puri P. Intestinal neuronal dysplasia. *Semin Pediatr Surg* 2003;12: 259-264
  5. Montedonico S, Acevedo S, Fadda B. Clinical aspect of intestinal neuronal dysplasia. *J Pediatr Surg* 2002;37:1772-1774
  6. Hatano M, Aoki T, Dezawa M, Yusa S, Iitsuka Y, Koseki H, et al. A novel pathogenesis of megacolon in Ncx/Hox11L.1 deficient mice. *J Clin Invest* 1997;15:795-801
  7. Von Boyen GB, Krammer HJ, Suss A, Dembowski C, Ehrenreich H, Wedel T. Abnormalities of the enteric nervous system in heterozygous endothelin B receptor deficient (spotting lethal) rats resembling intestinal neuronal dysplasia. *Gut* 2002;51:414-419
  8. Kanamori Y, Hashizume K, Sugiyama M, Tomonaga T, Terawaki

- K, Osawa K. Type B intestinal neuronal dysplasia. *Pediatr Int* 2005;47:338-340
9. . A case of intestinal neuronal dysplasia. *J Korean Pediatric Soc* 1996;39:1151-1157
10. Oguzkurt P, Senocak ME, Akcoren Z, Buyukpamukcu N. Diagnostic difficulties in neuronal intestinal dysplasia and segmental colitis. *J Pediatr Surg* 2000;35:519-521

J Korean Radiol Soc 2006;55:295 - 298

## Neuronal Intestinal Dysplasia in an Infant: Case Report<sup>1</sup>

Se Kyong Lim, M.D., Jong Chul Kim, M.D., Chung Dae Yoon, M.D.,  
Ji young Sul, M.D.<sup>2</sup>, Kyu Sang Song, M.D.<sup>3</sup>

<sup>1</sup>Department of Diagnostic Radiology, Chungnam National University Hospital

<sup>2</sup>Department of General Surgery, Chungnam National University Hospital

<sup>3</sup>Department of Anatomic Pathology, Chungnam National University Hospital

Neuronal intestinal dysplasia in pediatric patients has similar clinical symptoms and often similar radiologic findings to those of Hirschsprung's disease. Yet neuronal intestinal dysplasia shows hyperplasia of the myenteric plexus for the pathology, and it requires different treatment compared with Hirschsprung disease. This disease has been reported many times in Europe but, to date, only one case has been reported in the radiologic literatures in Korea. We report here on a case of neuronal intestinal dysplasia that involved the entire colon in a two-month-old boy, and we include the radiographic findings.

**Index words :** Infants, newborn, gastrointestinal tract  
Intestines, radiography

Address reprint requests to : Jong Chul Kim, M.D., Department of Diagnostic Radiology, Chungnam National University Hospital,  
640 Daesa-dong, Jung-ku, Daejeon, 301-721, Korea  
Tel. 82-42-220-7835 Fax. 82-42-253-0061 E-mail: jckim@cnu.ac.kr