Arch Jpn Chir 60(1), 75～79, Jan., 1991

症　　例

Two Cases of Bizarre Rib Anomalies Associated with Intestinal Malformation

Tsuneo Chiba, Takamichi Kamiyama and Ryoji Ohi

Deparment of Pediatric Surgery, Tohoku University School of Medicine
Received for Publication, Oct. 5, 1990.

Summary

Two cases of bizarre skeletal anomalies associated with intestinal malformation are presented. One patient had ileal atresia and Hirschsprung's disease, and the other anorectal malformation. Abnormalities in the number and morphology of ribs and vertebrae were present. Esophageal atresia often is associated with skeletal abnormalities, including supernumerary ribs, but these usually are not severe. Despite their severe costal malformations, neither patient had any complication after surgery to correct their intestinal malformation.

Introduction

FUNNEL CHEST and pigeon breast are common rib anomalies in children. Abnormalities in the number of ribs and severe morphologic deformities also have been reported[1], and rib anomalies occasionally are included in clinical syndromes[2,3]. In spite of this, there have been few reports on rib anomalies accompanying congenital diseases of the digestive organs. The present report describes two patients with a congenital anomaly of the intestinal tract associated with bizarre rib anomalies treated at our hospital and discusses rib anomalies other than funnel chest and pigeon breast in children with congenital malformations.

Case Report

Case 1

IY, a male infant, was born on May 22, 1985 weighing 3,430 g. Bilous vomiting and abdominal distension were noted immediately after birth. The patient was diagnosed as having congenital ileal atresia and underwent laparotomy the same day. The ileocecal region was excised, and ileocolostomy was performed for type III intestinal atresia. However, the patient was unable to defecate and a perforation of the ileum developed proximal to the anastomosis, necessitating ileostomy. The patient was suspected of having Hirschsprung's disease and was referred to our

Key words: Rib anomaly, rib deformities
索引語：肋骨奇形，肋骨変形.
Present address: Department of Pediatric Surgery, Tohoku University School of Medicine, 1-1 Seiryocho, Aobaku, Sendai 980, Japan.

hospital. After a complete examination the patient was diagnosed as having aganglionosis of the entire colon, and radical surgery (ileo-rectostomy with triangular flap method) was performed on July 12, 1986. This patient also had an unusual skeletal abnormality, with only 7 ribs on the left side, defects of the first through fifth ribs, and fusion of the 7th and 8th ribs (Fig. 1). Defects of the first to 7th thoracic vertebrae were present on the left side. No abnormality or muscular defect of the chest wall was present. The patient is now 4 years and 10 months of age and shows no hernia of the lung or respiratory problems.

Case 2

AA, a female, was born on May 6, 1988 weighing 2,370 g. An anovestibular fistula with imperforate fistula, torticollis and agenesis of the left kidney were present, and the patient was admitted to our department the day after birth. The fistula was dilated, and when the patient was 7 months old, an anal transposition procedure was performed. There were only 5 cervical vertebrae, abnormalities of the thoracic vertebral bodies (separation and fusion) and fused sacrum (Fig. 2). Scoliosis also was present. There were 13 ribs on the right side and 10 on the left. The patient continues to be followed and shows no abnormality of the chest wall.

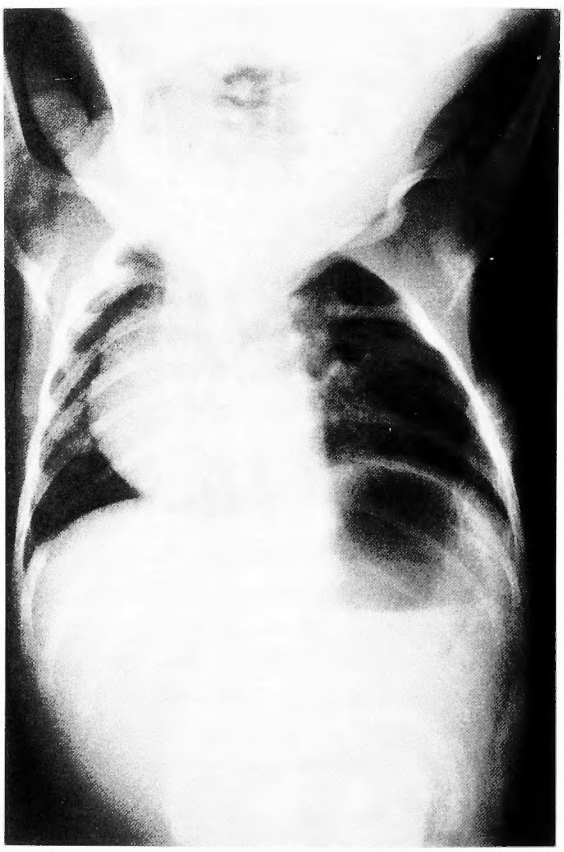

Fig. 1 Chest radiograph of case 1.

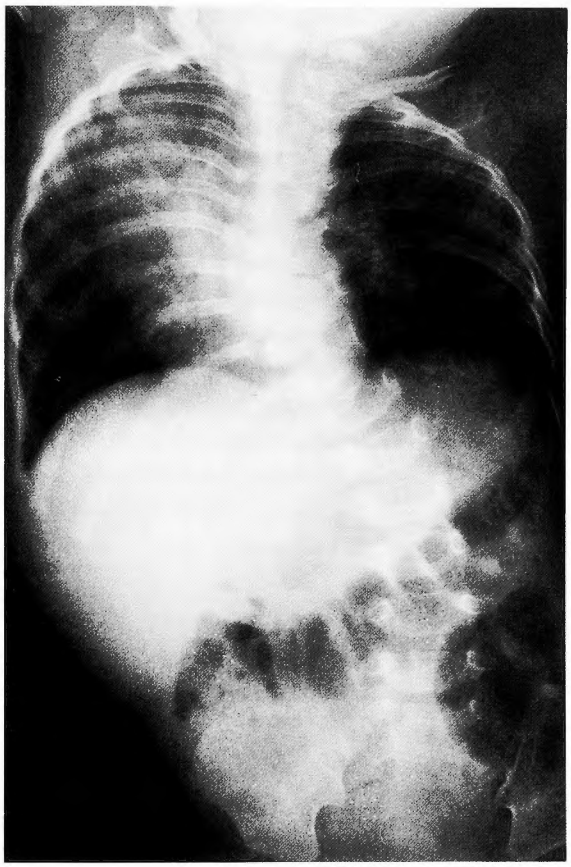

Fig. 2 Plain x-ray film of case 2.

Discussion

Congenital intestinal malformations often are associated with skeletal abnormalities. Of 3,378 patients admitted to Tohoku University Hospital since 1967, 66 (1.9%) had skeletal abnormalities

Table 1 Skeletal anomalies in pediatric surgical patients

	Total*	Skeletal anomaly**				
		Vertebral	Rib	Phalanx	Skull	Others
Esophageal atresia (56 cases)	22 (39.3%)	7	8	8	2	1
Anorectal malformation (214)	32 (15.0)	16	6	10	0	4
Abdominal wall defect (76)	4 (5.2)	2	0	0	0	3
Intestinal atresia (81)	4 (4.9)	4	2	0	0	0
Diaphragmatic hernia (68)	1 (1.5)	0	0	1	0	0
Biliary atresia (259)	2 (0.8)	0	0	0	2	0
Hirschsprung's disease (169)	1 (0.6)	1	1	0	0	0

* Number of the patients who have skeletal anomaly.
** If the patient have several anomalies, each anomaly calculated separately.

other than funnel chest, pigeon breast, and postoperative deformities. These skeletal abnormalities frequently occur concomitantly with congenital esophageal atresia and anorectal malformations (Table 1). The incidence of congenital esophageal atresia associated with skeletal abnormalities was highest, accounting for about 40% of cases, followed by anorectal malformation, abdominal wall defect, and intestinal atresia, in that order. The so-called VATER association was observed in 15 patients. The incidence of rib abnormalities in patients with esophageal atresia, anorectal malformation and intestinal atresia was 14%, 2.8% and 2.5%, respectively.

Rib anomaly may be manifested as an abnormal rib number or morphology. In esophageal atresia, 13 ribs often are present, and when number of vertebrae is increased, this condition may cause tension after surgery. Congenital malformation with an abnormality in the number of ribs include Poland's syndrome[2] and incontinentia pigmenti syndrome[3], but these are uncommon and most are not associated with an anomaly of the digestive tract. On the other hand, Jeune's disease[5] and severe costal malformation (reported as bizarre deformities by Ravitch[6]) have been reported as cases of costal deformities with absent ribs. Almost none of these cases were associated with an anomaly of the digestive tract. Our patients are believed to have suffered from a disease similar to that reported by Ravitch, but both patients had an associated congenital anomaly of the digestive tract. Surgical correction of severe chest wall deformities has been reported[1,7-10]. Neither patient in the present report underwent surgery to correct their costal anomaly because they were asymptomatic. However, thoracoplasty may become necessary to correct these severe deformities and scoliosis in the near future.

References

1) Ravitch MM: Atypical deformities of the chest wall—absence and deformities of the ribs and costal cartilages. Surgery **59**: 438–449, 1966.
2) Ravitch MM: Poland's syndrome. Congenital Deformities of the Chest Wall and their Operative Correction, WB Saunders, Philadelphia, 233, 1977.
3) Jones KL: Incontinentia pigmenti syndrome, Smith's Recognizable Patterns of Human Malformation, WB Saunders, Philaderphia, 448–449, 1988.
4) Touloukian RJ, Keller MS: High proximal pouch esophageal atresia with vertebral, rib and sternal anomalies: An additional component to the VATER association. J Pediatr Surg **23**: 76–79, 1988.
5) Jeune M, Beraud C, Carror R: Dystrophie thoracique asphyxiante de caractire familial. Arch Franc Pediatr **12**: 886, 1955.
6) Ravitch MM: Bizarre rib deformities, Pediatric Surgery, Year Book Med Pub, Chicago & London 583–584, 1986.
7) Ravitch MM: Asymmetric congenital deformity of the ribs, collapse of the right side to the chest. Ann Surg **191**: 534–538, 1980.
8) Hecker WCh & Daum R: Chirurgisches Vorgehen bei kongenitalen Brustwanddefecten. Der Chirurg. **35**: 482–484, 1964.
9) Ravitch MM: Congenital deformities of the chest wall and their operative correction. WB Saunders, Philadelphia 290–302, 1977.
10) Loeff DS, Creenwald MZ, Young SC, et al: A unique chest wall splint for unilateral agenesis of ribs. J Pediatr Surg **22**: 1129–1131, 1987.

和文抄録

腸管奇形を伴った奇妙な肋骨奇形の2例

東北大学小児外科
千葉　庸夫，神山　隆道，大井　龍司

肋骨奇形としては漏斗胸や鳩胸が日常しばしば見られ，また肋骨の数の異常として1，2本多い，あるいは少ないといった奇形が見られるが，中には数本以上欠損していたり，その他の椎骨の欠損を伴い異常な形を示すものがある．このような場合変形のみならず，肺のヘルニアをおこしたり，呼吸困難が生ずるため外科的な治療を必要とする事もある．これまでの報告では消化管の合併奇形を伴った例はほとんどない．われわれは回腸閉鎖症およびヒルシュスプルング病，低位鎖肛にともなった奇妙な肋骨奇形の症例を経験したので報告した．