Pediatr Radiol (1999) 29: 530–535

Marilyn J. Goske
John R. Goldblum
Kimberly E. Applegate
Craig S. Mitchell
Dianna Bardo

The "circle sign": a new sonographic sign of pneumatosis intestinalis – clinical, pathologic and experimental findings

Received: 21 September 1998
Accepted: 21 January 1999

Presented at the 41st Annual Meeting of the Society for Pediatric Radiology, Tucson, Arizona, May 1998

M. J. Goske (✉) · J. R. Goldblum · K. E. Applegate · C. S. Mitchell · D. Bardo
Section of Pediatric Radiology,
Cleveland Clinic Children's Hospital,
9500 Euclid Avenue, Cleveland,
OH 44195, USA

Abstract *Background.* Pneumatosis intestinals (PI) represents gas in the bowel wall. The appearance of PI using high-resolution ultrasound (HRUS) has not been well described.
Objective. The purpose of this report is to describe a new ultrasound sign of pneumatosis seen in three patients. This sign, called the "circle sign", is indicative of bubbles of gas within the circumference of the bowel, producing an appearance of a continuous echogenic ring on ultrasound. Further studies of the sonographic characteristics of pneumatosis were performed with an in vitro model.
Materials and methods. HRUS was performed prospectively in three patients demonstrating extensive PI radiographically. The appearance of the gas was characterized and the behavior of the intramural bubbles was studied when the bowel was compressed with the ultrasound transducer. Either CT scan or pathologic correlation was obtained in all patients. Experimental models of PI using air injected into the wall of sausage casing were developed.
Results. The presence of echogenic gas bubbles within the circumference of the wall of the bowel seen with HRUS was shown to represent pneumatosis intestinalis at histologic examination or by CT scanning in the three study patients. In vitro studies confirmed the clinical impression that the use of compression is helpful in distinguishing intramural from intraluminal air.
Conclusion. The presence of echogenic gas bubbles in the wall of the bowel, often seen as a circle within the circumference of the bowel, may be helpful in diagnosing PI on ultrasound using HRUS.

Introduction

Pneumatosis intestinalis (PI) represents gas in the bowel wall [1]. Although the precise pathogenesis of PI is obscure, mechanical or inflammatory disruption of the bowel wall with loss of mucosal integrity and subsequent bacterial invasion of the wall is thought to play a significant role [2].

The clinical significance of PI varies with the age of the patient and underlying disease state. In premature infants or those with complex congenital heart disease, the presence of PI often indicates serious morbidity. In fact, the presence of PI radiographically is the "single most important factor" in the staging of necrotizing enterocolitis (NEC) [2].

However, pneumatosis is difficult to diagnose on radiographs [3], and there is a high degree of interobserver disagreement as to its presence on the conventional radiograph [4]. It would be advantageous to have other objective tests to substantiate its presence because early detection and treatment of children with PI, particularly those with NEC, result in improved survival [5].

Vernachia and Jeffrey were the first to report the ultrasound recognition of PI in 1985 [6]. Since then, there have been sporadic reports describing the US features

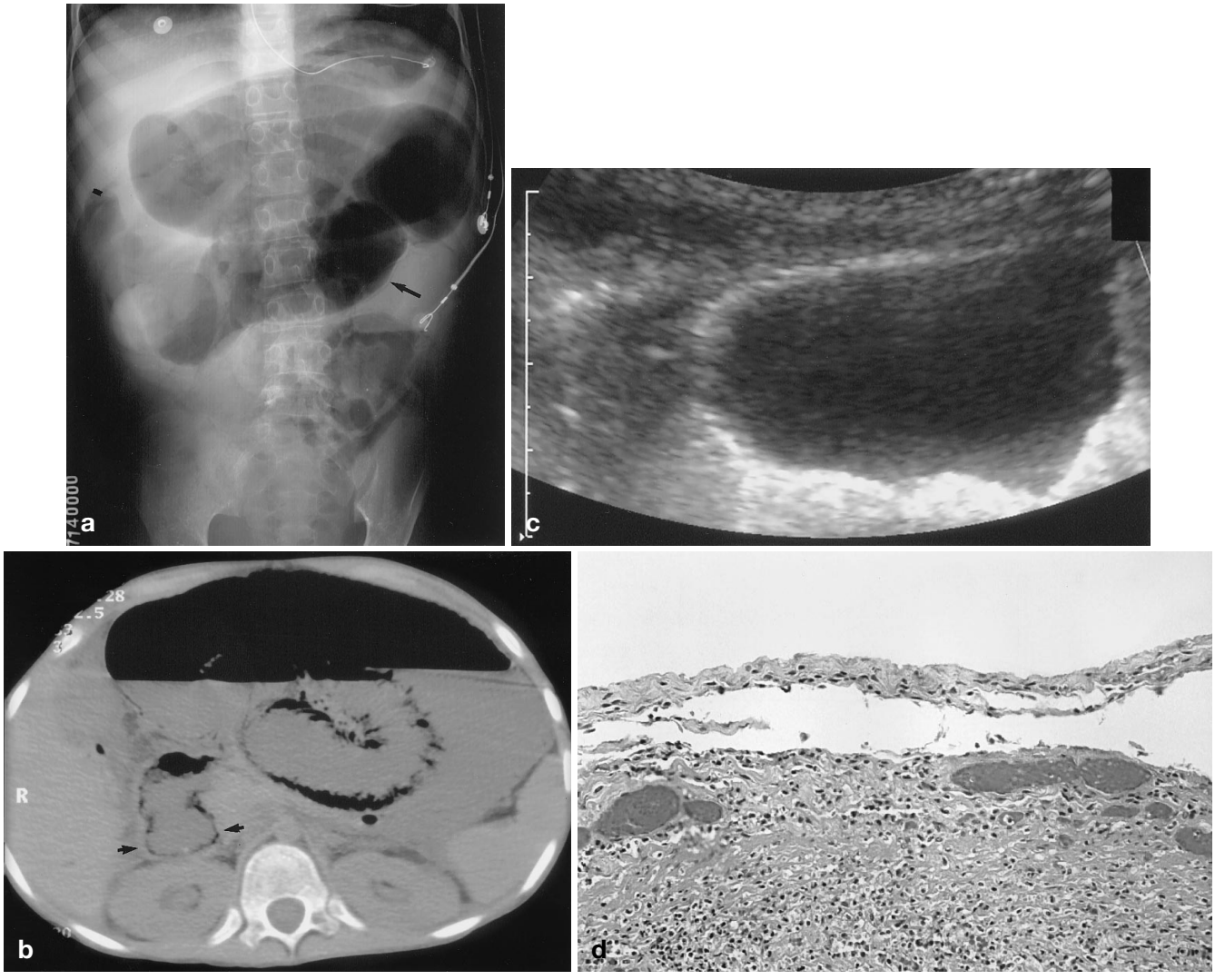

Fig. 1 **a** A 15-year-old boy with cerebral palsy with obstructed bowel from adhesions and pneumatosis. Plain radiograph demonstrates severe proximal small-bowel dilatation and linear pneumatosis intestinalis following the contour of the bowel wall *(arrow).* **b** CT demonstrates marked distension of proximal small-bowel loops with severe pneumatosis involving the circumference of the wall. In this scan, the bowel loops are not visualized in true cross section and only a partial "circle" of pneumatosis is seen *(arrows).* **c** US shows the "circle sign" with coarse echogenic gas seen within the circumference of the wall of the proximal small bowel. **d** Histologic specimen demonstrates coagulation necrosis compatible with ischemia. Large air spaces are secondary to pneumatosis in a subserosal location

of PI, only one of which to our knowledge has attempted to describe the patterns of PI in any detail [7].

Our study attempts to characterize further the behavior and appearance of PI using HRUS, histologic correlation and an in vitro model. One pattern, which we have termed the "circle sign", was seen in our three study patients, all of whom had florid pneumatosis radiographically. This pattern was recognized when extensive gas bubbles were seen in the entire circumference of the bowel wall. This sign and the appearance and behavior of PI when studied with HRUS are the subjects of this report.

Materials and methods

Three children with extensive pneumatosis identified radiographically were prospectively studied using high-frequency transducers (Siemens Ellegra, Siemens Medical System, Erlangen, Germany or HDI 3000, Advanced Technology Laboratories, Bothell, Washington, USA). A linear 7–10 MHz transducer was used to scan the bowel in a rasterlike fashion. The abdomen and pelvis were divided into 16 segments, and sagittal images were recorded. The liver was scanned for the presence of portal venous gas. The bowel was inspected for the presence of PI, wall thickening, and perito-

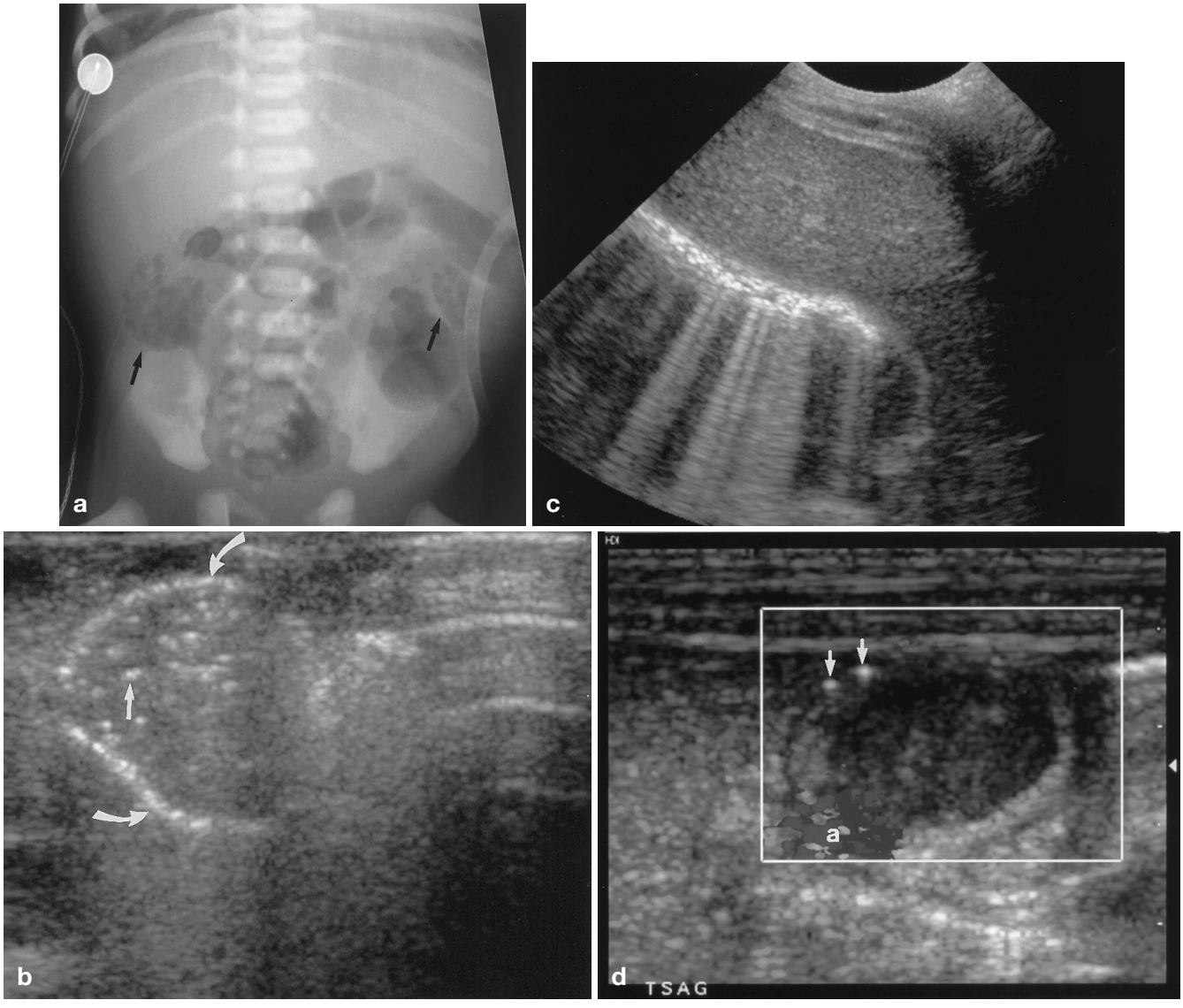

Fig. 2 **a** Full-term infant with DiGeorge's syndrome, truncus arteriosus, and pneumatosis intestinalis. Radiograph demonstrates a bubbly appearance in the right and left abdomen compatible with pneumatosis *(arrows)*. Bowel distension is also seen without evidence for portal venous gas. **b** High-resolution US during the patient's first episode of grossly bloody stool demonstrates the "circle sign" of pneumatosis intestinalis in a dilated loop of small bowel. The intraluminal gas bubbles *(straight arrow)* moved with compression, while the coarse bright echoes in the wall *(curved arrow)* did not. **c** Transverse US scan through the lower aspect of the liver demonstrates the "aurora sign" of pneumatosis. This shows alternating stripes of bright echoes separated by acoustic shadows and is seen only with a solid acoustic window such as liver. **d** Color US shown in black and white obtained 10 days after the resolution of the first episode of gross pneumatosis shows a very different appearance on US. On this examination, only two air bubbles *(arrows)* within the wall of a loop of small bowel were seen. These bubbles did not move with compression and were very different in appearance from intraluminal contents. No color flow was seen in the wall. Artifactual "color" was seen from flash artifact *(a)*. The cecum (not shown) was thick-walled

neal fluid. In areas where PI was suspected on ultrasound, compression of the anterior abdominal wall or changing of patient position was performed. This was done to observe changes in the echogenic bubbles relative to the bowel wall (i.e., intramural or intraluminal). Shortly thereafter, CT scanning or histologic confirmation of PI was obtained in all cases.

Case 1

A 15-year-old boy with a history of cerebral palsy and recently diagnosed Hirschsprung's disease presented with marked abdominal distension. The initial radiograph (Fig. 1 a) demonstrated marked proximal small-bowel distention and PI. The CT scan, performed at the request of the referring physician, within 30 min demonstrated marked distension of bowel loops, with PI seen circumferentially within the wall of the bowel (Fig. 1 b). Minimal portal venous gas was seen. Ultrasound performed immediately after CT demonstrated portal venous gas as well as echogenic foci within the wall of the dilated small bowel (Fig. 1 c). The intramural gas

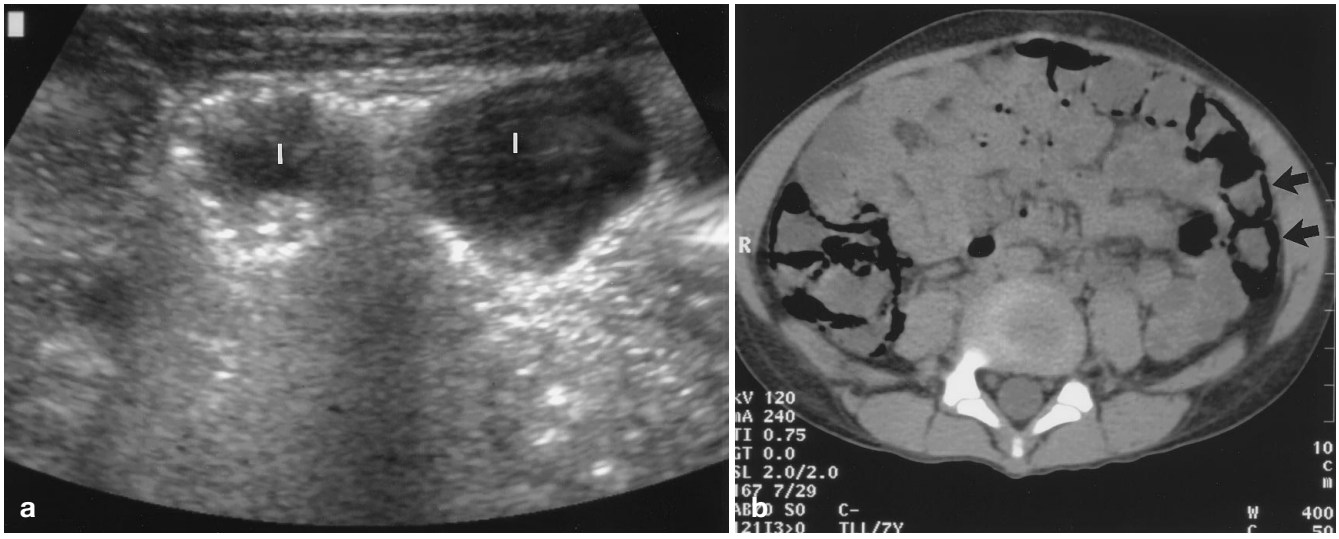

Fig. 3 **a** US demonstrates course echogenic foci within the walls of two adjacent loops of bowel. The "circle sign" is seen (*l* loop). **b** CT scan confirms extensive pneumatosis, some in a circular pattern *(arrows)* within the bowel wall, as was seen on US and the radiograph

was brighter and coarser than intraluminal gas and did not move with compression of the anterior abdominal wall or decubitus positioning of the patient. In several loops, the air remained constant in a circular distribution. No wall thickening or ascites was seen. Twenty minutes after completion of the ultrasound, as the patient was being prepared for surgery, the patient died, presumably from air embolism. At autopsy, dilated ischemic bowel with obstruction from adhesions was seen. The bowel was diffusely crepitant, indicating the presence of PI. A histologic evaluation of bowel sections thought to contain PI on HRUS was done. These specimens demonstrated clear spaces of varying size within the bowel wall, predominantly in a subserosal location. Although scattered giant cells surrounded those spaces, there was no significant inflammatory response, fluid, or hemorrhage in the wall. The spaces were not lined by cells. This indicates that they are air-filled spaces as opposed to vascular spaces lined with endothelial cells (Fig. 1 d).

Case 2

This full-term male infant was found to have DiGeorge's syndrome with truncus arteriosus. At 10 days of age, the patient developed blood in his stools. Radiography demonstrated a bubbly pattern of gas compatible with PI (Fig. 2 a). HRUS performed within 30 min demonstrated portal venous gas (not shown) and PI in the bowel wall in some loops in a circular pattern (Fig. 2 b). A second HRUS, performed 10 days later when the patient had again deteriorated, demonstrated the "aurora" sign of pneumatosis (alternating bands of echogenicity) [7] (Fig. 2 c) and PI in the bowel wall. This could be particularly well seen near the cecum and terminal ileum, which was marked by the appendix within peritoneal fluid (Fig. 2 d). Within 2 h, the patient went to the operating room and underwent bowel resection. Histology of the resected bowel of the terminal ileum and cecum revealed clear spaces of varying size in a submucosal location, consistent with pneumatosis.

Case 3

A 7-year-old boy with combined immune deficiency developed abdominal pain and distention. Stool guiac was positive. Radiograph (not shown) showed extensive PI in the ascending and descending colon. HRUS performed within 2 h demonstrated PI in a circular pattern in the colon, which stayed within the wall when the abdominal wall was compressed (Fig. 3 a). No bowel-wall thickening, ascites, or portal venous gas was seen. A CT scan performed immediately afterward (Fig. 3 b) confirmed the distribution and patterns of the pneumatosis seen on HRUS.

The second component of this study was to use an in vitro model to better define the behavior of the intramural gas bubbles. A model of the bowel was made using a double layer of sausage casing (length of 6 in.), which was filled with either water or a thick liquid-food substitute (Ensure, Ross Laboratory, Columbus, Ohio), tied at both ends and immersed in a water bath. The sausage casing was filled with two different materials to simulate different bowel contents. Next, 0.3–0.55 cc of air was injected between the two layers of casing using a 25-gauge needle and a tuberculin syringe. Ultrasound images were obtained before and after air injection and manual compression to the casing while in the water bath. Observations as to the ability to detect the bubbles and the movement of the bubbles in the wall and in the luminal contents were made.

Results

The histology results from two patients confirmed that pneumatosis was indeed present in the loops of bowel thought to have PI on HRUS. There was no other material in the wall, such as fat or hemorrhage, that could have produced the echogenic foci seen on HRUS. Care was taken to correlate precise bowel loops from the HRUS with the histology.

In the sausage-casing model, intraluminal gas in water moved obviously and easily with compression (Fig. 4 a). Intraluminal gas in the thicker liquid was more difficult to distinguish from intramural air due to less bubble motion (Fig. 4 b). Repeated compression

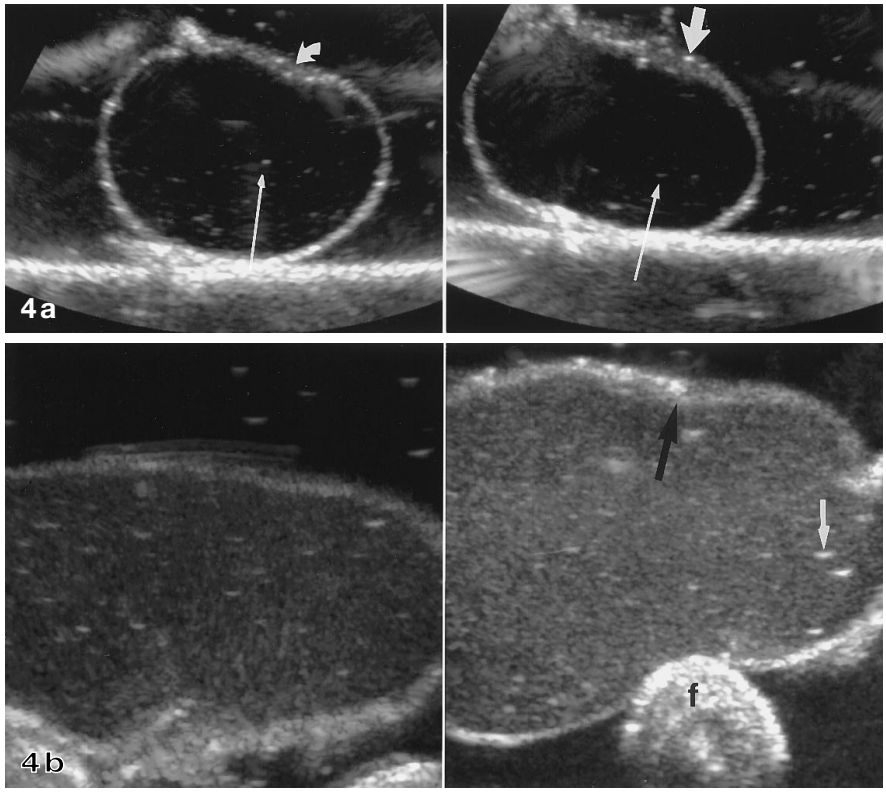

Fig. 4 **a** An "in vitro" model using two layers of sausage casing filled with water immersed in a water bath. The image on the left, without compression, demonstrates air *(curved arrow),* which has been injected into the "wall". With compression, the air remained within the wall *(straight arrow).* The intraluminal air bubbles *(arrowhead)* as well as air bubbles in the water bath *(white arrow)* moved on real-time inspection with only a minor degree of manual compression. **b** An "in vitro" model using two layers of sausage casing filled with Ensure immersed in a water bath. Image at left is baseline, with no air injected into the wall and no compression. Low-level echoes within the lumen represent the thicker liquid, while the bright echoes, which slowly moved, were related to settling of bubbles after immersion in the water bath. Image on the right, after air was injected into the wall *(black arrow)* using manual compression (*f* fingertip) shows bright echoes, which remained stationary, simulating pneumatosis. The intraluminal bubbles were observed as moving echoes within the thicker liquid *(white arrow)*

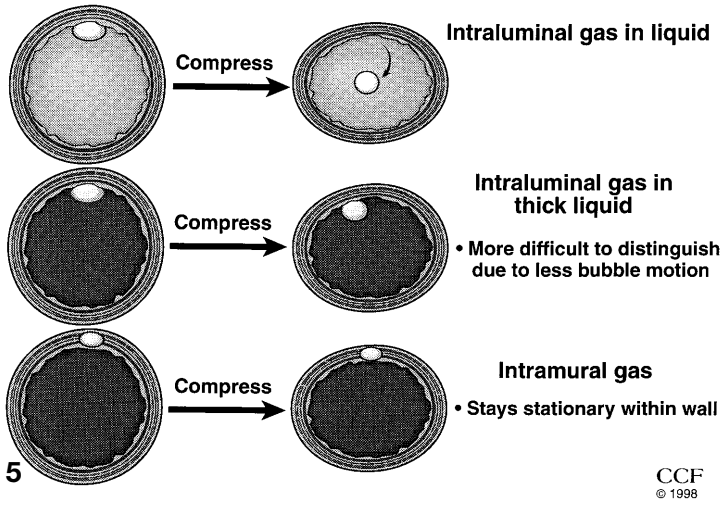

Fig. 5 Illustration summarizing clinical and "in vitro" experiments of the behavior of intraluminal gas and intramural gas in thin and thick liquids with compression. In the thicker liquid, the bubbles in the lumen may be more difficult to observe. The intramural gas as is seen in pneumatosis stays stationary within the wall, even with compression

caused further production of intraluminal bubbles, which was helpful in defining the true margins of the lumen. In this in vitro situation, the bubbles could be displaced from the periphery of the lumen, thereby more readily distinguishing the intraluminal from intramural gas. With both liquids, air injected into the wall did not move with compression, but was more difficult to detect in the thicker liquid. The ultrasonographer is relying on several features to detect PI: actual visualization of the gas in the wall, lack of motion of the bubbles over time in the periphery of the bowel wall, and lack of movement of the bubbles with compression. As bowel contents thicken, it appears as though confidence in the detection of PI may decrease because the wall may be more difficult to distinguish and bubble motion is less obvious (Fig. 5).

Discussion

The etiology of PI in children is diverse and still not well understood. Subtotal ischemia of the intestine with progressive devitalization of tissues may result in bacterial invasion and inflammation. Mechanical or inflammatory disruption of the bowel wall is thought to play a significant role [1]. In experiments in piglets, pneumatosis appears to originate in the lymphatic vessels within the submucosa. Stagnant material in distended lymphatics may provide a substrate for the production of hydrogen gas by proliferating anaerobes producing the gas seen on the radiograph, although this is speculative [2]. The location of pneumatosis is most often submucosal, although it may be transmural in 22 % of patients [8].

To date, there has been little discussion of the appearance of PI as seen on ultrasound examination of the bowel. In 1985, Vernachia and Jeffrey [6] described a single adult patient with pneumatosis detected on ultrasound before it was observed on the radiograph. Two articles by Bomelburg et al. [5, 9] discuss the visualization of pneumatosis and the use of ultrasound as a more sensitive method than radiographs in detecting this condition in premature infants with necrotizing enterocolitis. Kohzaki et al. [7] discuss the "aurora sign", an ultrasound sign of pneumatosis cystoides intestinalis in an adult patient, a pattern seen in patient 2. They describe long and short stripes of bright echoes separated by acoustic shadows seen emanating from the bowel wall when the liver was used as an acoustic window. They were able to reproduce these findings with an experimental model.

In this report, we attempt to characterize the appearance and behavior of PI using HRUS. In our experience with patients as well as the in vitro model, the detection of PI is most difficult in situations where there is minimal fluid-filled bowel contents, collapsed bowel, lack of peristalsis or movement of luminal contents, and, lastly, more solid bowel contents with air bubbles trapped within. The "circle sign" was helpful in drawing our eye towards the more obviously diseased loops of bowel for further observation. It increased our confidence that PI was present, as one would expect gas to rise to the highest point within the lumen if it were intraluminal. In the "circle sign", portions of gas are trapped dependently. In some patients, only occasional gas bubbles are seen scattered throughout or in portions of the bowel wall. These minute quantities of gas may be seen to be within the wall using HRUS. In our experience, the bubbles do not always produce a "dirty" shadow, but in some instances are seen as intramural. The presence of ascites appears to enhance the ability to detect PI by providing clearer definition of the outer bowel surface.

Conclusion

This study was limited to only three patients with florid pneumatosis, and further study with this technique will be necessary to determine if smaller quantities of PI will be detectable. In addition, analysis of the impact of this imaging technique, if any, on physician practice patterns will be necessary. However, these preliminary results suggest that HRUS may provide an additional method of assessing high-risk patients for the presence of PI.

In our study, pneumatosis intestinalis appears to be better seen with the advent of high-frequency transducers. In some cases, bubbles of gas trapped in an intramural location may be able to be differentiated from intraluminal bubbles by their lack of motion with compression or peristalsis. Furthermore, the presence of gas seen within the circumference of the wall of the bowel, the "circle sign", was helpful in our three patients with extensive pneumatosis.

The results of this study should be confirmed in a prospective, multicenter trial to determine whether this method has promise in a larger patient population at different sites and in patients with smaller amounts of PI.

References

1. Wood BJ, Kumar PN, Cooper C, et al (1995) Pneumatosis intestinalis in adults with AIDS: clinical significance and imaging findings. AJR 165: 1387–1390
2. Sibbons P, Spitz L, Van Velzen D (1992) The role of lymphatics in the pathogenesis of pneumatosis in experimental bowel ischemia. J Pediatr Surg 27: 339–342
3. Rencken IO, Sola AA, Al-Ali F, et al (1997) Necrotizing enterocolitis: diagnosis with CT examination of urine after enteral administration of iodinated water-soluble contrast material. Radiology 205: 87–90
4. Mata AG, Rosengert RM (1980) Variability in the radiographic diagnosis of necrotizing enterocolitis. Pediatrics 66: 68–71
5. Bomelburg T, von Lengerke HJ (1992) Sonographic finding in infants with suspected necrotizing enterocolitis. Eur J Radiol 15: 237–240
6. Vernachia FS, Jeffrey RB (1985) Sonographic recognition of pneumatosis intestinalis. Am J Roentgenol 145: 51–52
7. Kohzaki S, Hayashi K, Fukuda T, et al (1994) Case report: The "aurora sign" – a new sonographic sign of pneumatosis cystoides intestinalis. Br J Radiol 67: 1275–1277
8. Ballance WA, Dahms BB, Shenker N, et al (1992) Pathology of neonatal necrotizing enterocolitis: a ten-year experience. J Pediatr (Suppl) 117: 6–13
9. Bomelburg T, von Lengerke HJ (1992) Intrahepatic and portal venous gas detected by ultrasonography. Gastrointest Radiol 17: 237–240