

Gas in the Intestinal Wall and Portal Venous System in Infants

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The presence of gas in the intrahepatic portal veins can be recognized on the plain roentgenogram of the abdomen. This radiological finding was first described by Wolfe and Evans in 1955.¹ Since all six infants in their series died a short time after portal venous gas was detected, a grave prognostic significance of forecasting impending death was ascribed to this sign. It is frequently accompanied by gas in the intestinal wall producing a typical ring-like radiolucency around the gas is the bowel lumen.^{2 3}

A total of forty three pediatric cases with portal venous gas has been reported in the English literature.⁴ Its occurrence could not be as rare as the number of reported cases would suggest since we have observed seven infants with this entity in a 20 month period. Together with two other infants we had studied in earlier years, we wish to present our experience with nine patients with portal venous gas and compare our findings with those reported in the literature.

Report of Cases

The pertinent clinical features of our patients with portal venous gas are outlined in Table I. It is interesting to note that all the infants are less than six months of age. Five patients are premature babies.

Clinically these nine infants fall into two groups:

1. In the first group there are seven patients with gastroenteritis (Cases 1 to 7). Two of these had bacteriologically proven sepsis as well (Cases 2 and 6). The pathogenesis of the portal gas is probably asso-

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ciated with their gastroenteritis and every single one of these seven infants died within two days following demonstration of gas in their portal veins.

2. In the second group there are two patients (Cases 8 and 9) who had no evidence of gastroenteritis. One of these infants had an umbilical catheter and had two exchange transfusions. Gas may have been introduced into the portal system through this catheter. The second child had a congenital anal stenosis and underwent a series of progressive dilatations. Both of these patients survived.

The radiological findings in our patients are shown in Figures 1 to 9 and are analyzed in Table II. Marked gaseous distention of the bowel and air-fluid levels on the upright radiograms are seen to be universally present in association with gas in portal veins. Only a single patient in our series (Case 2) had pneumoperitoneum indicating perforation. Seven infants out of the total nine had gas in the intestinal wall in addition to portal venous gas, indicating a close relationship between these two findings.

Post-mortem examinations were done on two of the patients who died (Cases 2 and 3). Both showed multiple gas containing cysts in intestinal wall (Figure 10). Case 2 also had mucosal ulcerations with pseudomembrane formation and diffuse inflammatory reaction consistent with necrotizing enterocolitis. This infant also had truncus arteriosus malformation in the heart, focal hepatic necrosis and tubular necrosis in the kidneys.

Discussion

The presence of gas in the portal venous system must be differentiated from gas in the biliary tree since the prognosis in these two types of patients is entirely different. The latter may be the result of the incompetence of the sphincter of oddi in the newborn⁵ or a fistulous connection between the biliary system and the bowel in later years and does not necessarily indicate a fatal outcome. Fortunately these two types of gas collection within the hepatic shadow can be easily differentiated radiologically. Gas in the portal veins has a characteristic branching pattern, radiating from the porta hepatis to within 2 cm. of the hepatic margin.^{6 7 8} In contrast gas in the bile ducts is centrally located where it accumulates in the major bile ducts at the porta hepatis and/or the gallbladder. This difference in the distribution of the gas is apparently due to the cephalad flow in portal veins versus the caudal flow of bile.

TABLE I
CLINICAL FINDINGS OF NINE PATIENTS WITH PORTAL VENOUS GAS

Patient	Age	Sex	Birth Weight	Gestation	Gastro-enteritis	Sepsis	Stool Culture	Congenital Anomalies	Fatal Outcome
1. A.D. 202682	6 days	M	1800 gm.	8 months	+	—	E. Coli Agg (—)	—	+
2. T.A. 299792	21 days	M	2450 gm.	?	+	+	E. Coli Agg (—) E. Coli Occult Blood	Truncus arteriosus, V. S. D.	+
3. C.G. 68/63337	4,5 months	M	—	—	+	—	E. Coli Agg (+) Fresh blood	—	+
4. K.B. 214012	9 days	M	1000 gm.	28 weeks	+	—	—	—	+
5. D.H. 299207	5 months	M	—	—	+	—	E. Coli Agg (—)	—	+
6. E.K. 308434	17 days	F	1710 gm.	30 weeks	+	+	E. Coli Agg (—) A. aerogenes Pyocyaneus	—	+
7. T.Ç. 282642	5 months	M	—	—	+	—	E. Coli Agg (—)	—	+
8. B.B. 206501	6 days	F	1610 gm.	35 weeks	—	—	E. Coli Agg (—)	—	Survived Discharged on the 50 th day.
9. H.D. 66/35754	4 days	M	—	—	—	—	—	Anal stenosis	Survived Discharged on the 11 th day.

TABLE II
ANALYSIS OF RADIOLOGICAL FINDINGS IN OUR PATIENTS WITH PORTAL VENOUS GAS

Patient	Gas in portal veins	Gas in intestinal wall	Pneumoperitoneum	Gaseous Distention	Air-fluid levels	Time of X-ray before death
1. A.D. 202682	+	+	—	+	+	2 hours
2. T.A. 299792	+	+	+	+	— (Supine X-ray)	20 hours
3. C.G. 68/63337	+	+	—	+	+	1 day
4. K.B. 214012	+	—	—	+	+	2 days
5. D.H. 299207	+	+	—	+	— (Supine X-ray)	2 days
6. E.K. 308434	+	+	—	+	+	1 day
7. T.Ç. 282642	+	—	—	+	+	36 hours
8. B.B. 206501	+	+	—	+	+	Patient survived
9. H.D. 66/35754	+	+	—	+	+	Patient survived

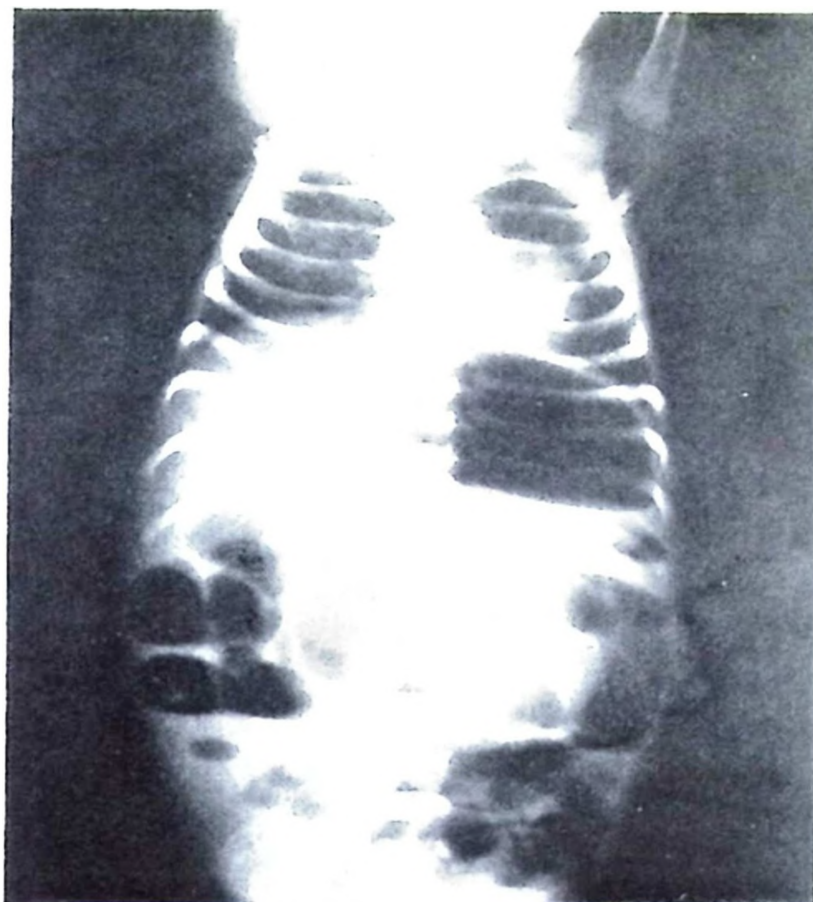


Figure 1

Figures 1 to 9 show the plain roentgenograms of the abdomen of Cases 1 to 9 respectively. Gas in portal veins is present in all of them.

A commonly associated finding in patients with portal venous gas is the presence of intestinal intramural gas collections, the so-called "Pneumatosis cystoides intestinalis."^{9 10 11} This was definitely present in 7 of our 9 patients. Since radiological demonstration of intramural gas is much more difficult than the demonstration of portal venous gas, this association may actually be more close i.e. almost 100%.

Another radiological finding that may also be present in these infants is free air in the peritoneal space due to perforation. Pneumoperitoneum was present in one of our patients.

The pathogenesis of portal venous gas is not definitely known. Two major hypotheses have been offered to explain the mechanism by which the gas enters the portal veins.^{12 13} One theory suggests that gas under increased tension within the lumen of the bowel will pass through its wall altered by impaired circulation due to severe distention or thrombosis. Thus it will move into mesenteric veins and into the portal venous system. The fact that in all the reported infants severe dilatation of the bowel loops was present supports this theory.¹⁴⁻¹⁶



Figure 2

A large amount of free air is present in the peritoneal space under the left hemidiaphragm in the radiogram of Case 2.

Severe gaseous distention of the bowel was readily recognizable in all our patients (Table II). On the other hand the many cases of bowel distention without portal vein gas indicate that other factors must play a part, predisposing the intestinal wall to the passage of intraluminal gas. Surgical exploration or post-mortem examinations of several patients with portal venous gas have disclosed loss of mucosal integrity either due to necrotizing enterocolitis or gangrene following mesenteric vein thrombosis.^{4 17} Mucosal destruction will range from single or multiple perforations to actual loss of large areas of mucosa.¹⁰

As a matter of fact, all cases of gas in the portal venous system in adults were associated with mesenteric thrombosis.¹⁶ Thus, alteration in the intestinal wall permeability due to vascular impairment may very well be the critical factor in allowing the passage of intraluminal gas into the mesenteric veins.

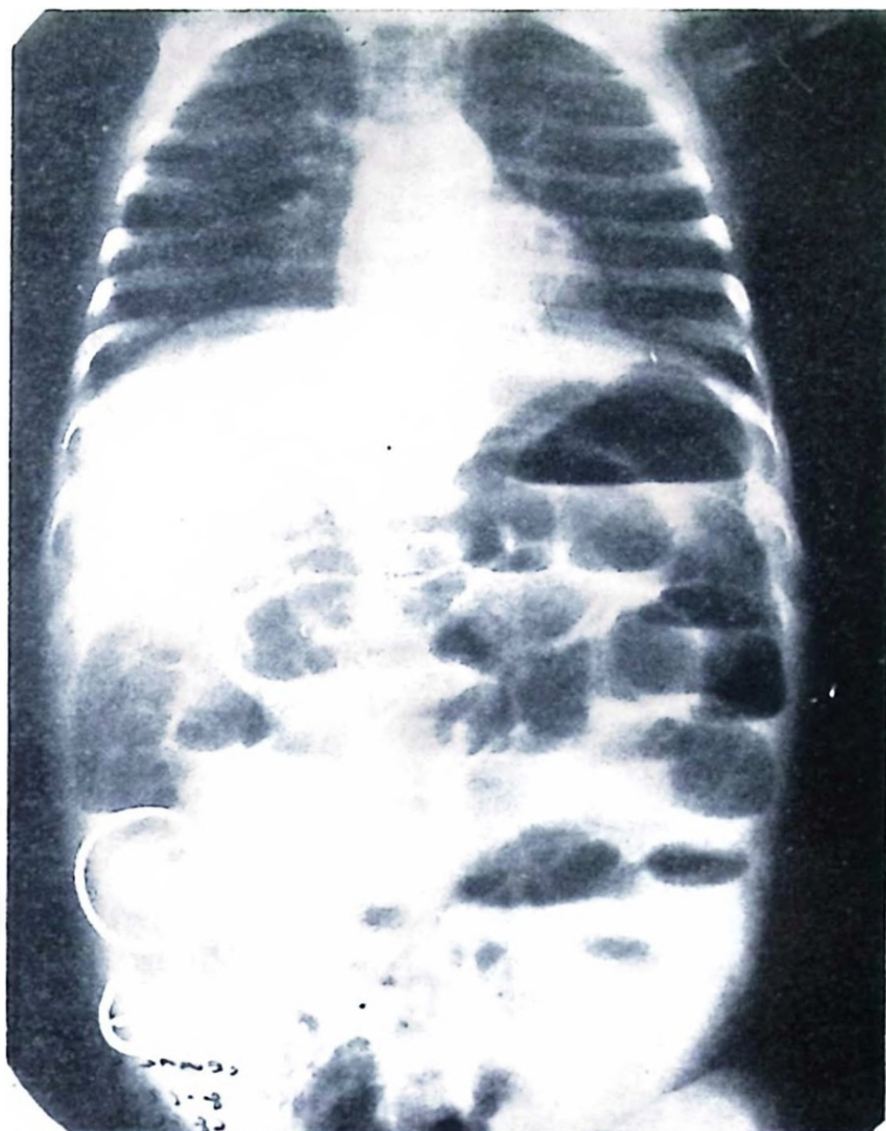


Figure 3

In children, if we exclude those cases where air was apparently introduced into the portal veins through umbilical catheterization^{2,8} or a peroxide enema¹⁸ a history of diarrhea and/or vomiting is present in all the reported cases of portal gas. The dehydration and the ensuing hemoconcentration of these infants might be a predisposing factor for the thrombosis of the mesenteric vessels. Actually microscopic sections of the intestinal mucosa of these affected infants have shown capillary thrombi and coagulation necrosis of mucosa without much inflammatory cellular response.¹⁰

Intestinal bleeding is not an uncommon finding in necrotizing enterocolitis and was present in two of our patients. If the bowel wall is damaged enough to allow the passage of blood into the lumen than one can speculate that gas in the lumen under sufficient pressure may pass into the capillaries through the same route. The amount of this

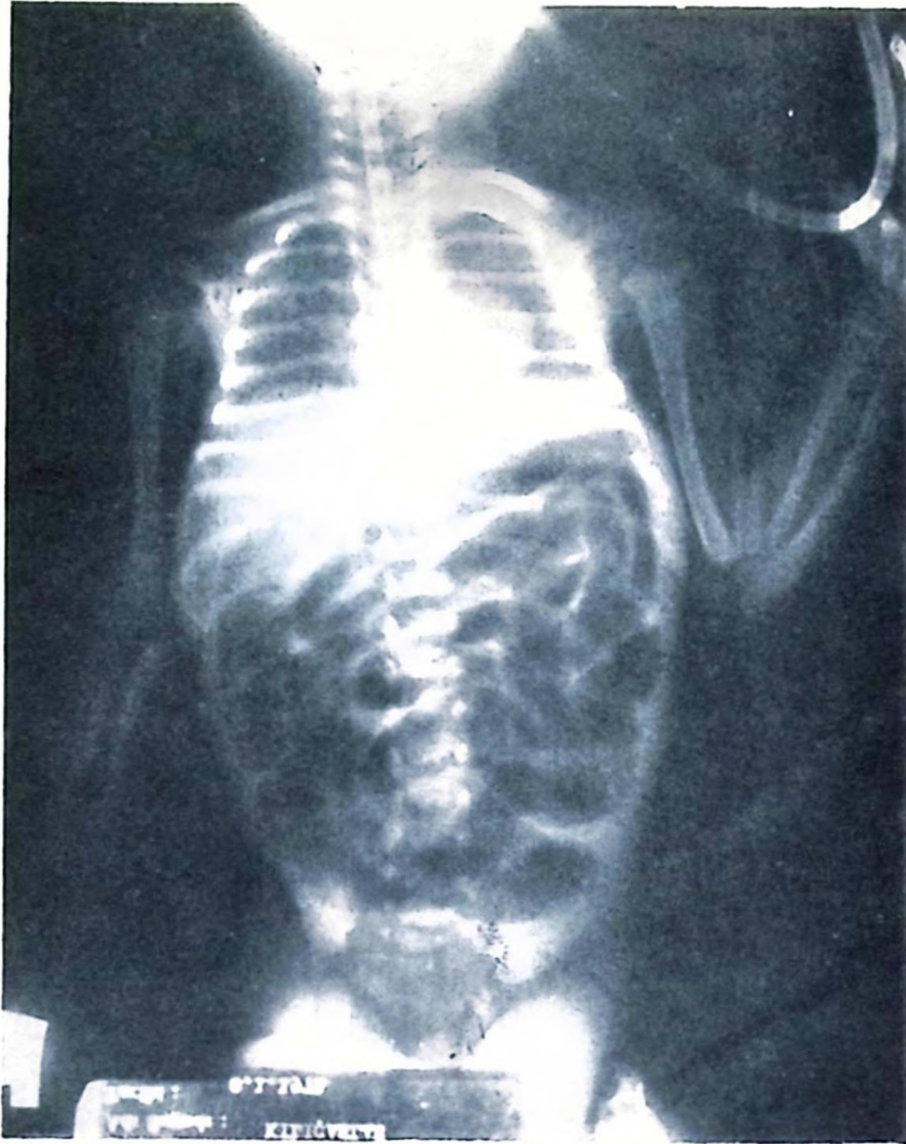


Figure 4

gas need not be great in order to be visualised in the portal veins since it has been demonstrated in the dog that as little as 5 c.c. of air injected into the mesenteric vein is sufficient to outline the portal system of the left lobe of the liver.⁶

The high incidence of intestinal intramural gas in patients with portal venous gas is also in support of this mechanical "passage through the wall" theory. Here one must differentiate the gas cysts in the bowel wall of these infants from the benign "pneumatosis cystoides intestinalis" seen in adults. The gas filled cysts under discussion here, do not have a cellular lining, may be submucosal as well as subserosal and are usually associated with extensive necrosis of tissue around them, all in contrast with the adult form.^{19 20} And again, since the presence of these cysts in an infant signals serious disease, whereas the adult



Figure 5



Figure 6



Figure 7

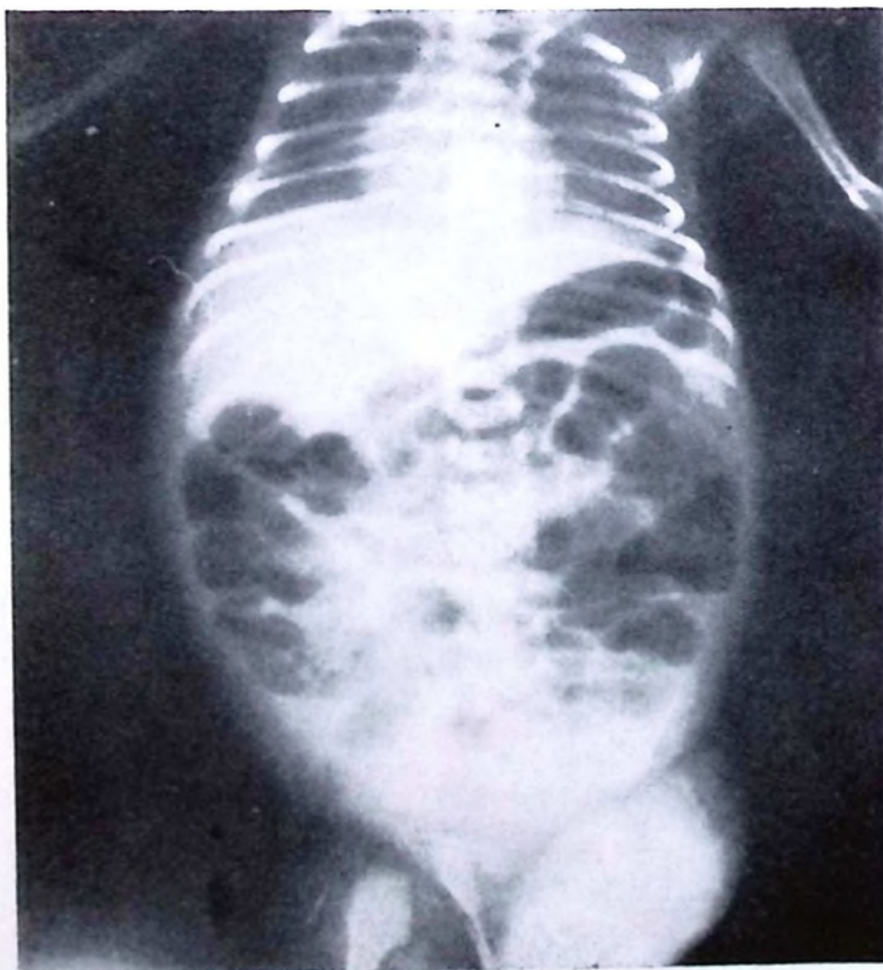


Figure 8



Figure 9

Intestinal intramural gas is most readily discernable in the radiogram of Case 9 (Figure 9) as crescent shaped radiolucent bands around intestinal loops.

form does not, the term "pneumatosis cystoides intestinalis" should not be used to describe these changes in children.

The second theory trying to explain the origin of the gas in the portal veins, suggests that the gas is produced by enteric organisms which have entered the vascular system through compromised bowel wall. Patients of Wiot and Felson²¹ had blood cultures positive for gas forming organisms. Infections probably do have a part in altering the permeability of the bowel wall, but many cases without sepsis (including seven cases of ours out of nine) and the absence of gas elsewhere in the vascular system in any of the reported cases speak against formation of the gas directly by the bacteria.

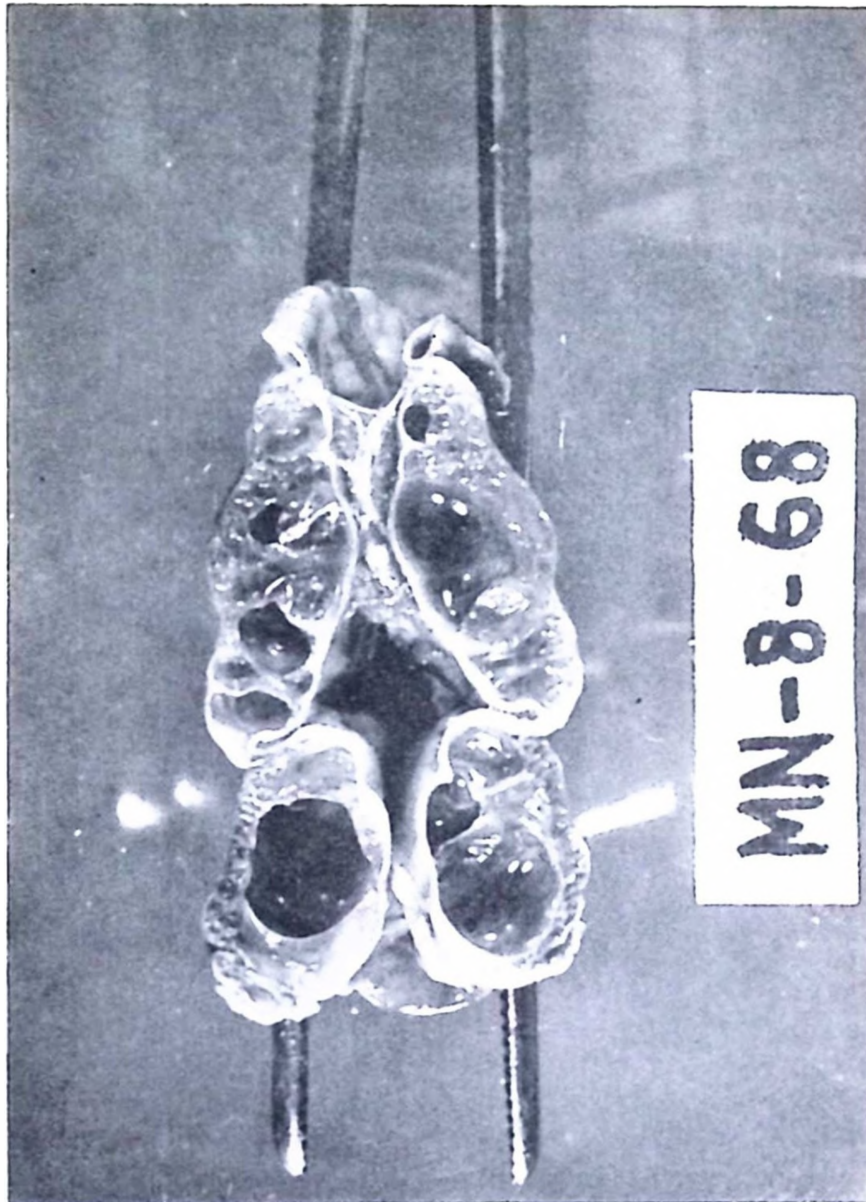


Figure 10

Cross section of the wall of a segment of ileum from Case 3, showing multiple intramural cysts of varying size.

Review of the literature shows that portal vein gas in children has been seen in the following conditions: necrotizing enterocolitis in premature infants, intestinal obstruction as in imperforate anus, duodenal atresia or aganglionosis, mesenteric vein thrombosis with gangrene of bowel and erythroblastosis fetalis. The cases where gas is artificially introduced through an umbilical catheter or a peroxide enema should not be included among this spectrum of diseases.

Whatever the underlying basic disease may be, air bubbles in the portal veins or the bowel wall always indicate the same pathology: that bowel is compromised. Especially when intramural gas is noted locally, it may indicate a surgically resectable localized lesion.

It is due to this understanding that, thanks to early surgery, some infants have survived in recent years regardless of the gas seen in their portal veins.¹⁷ The possibility of survival has made recognition of portal venous gas all the more important. One should always be alert for these abnormal gas collections in the intestinal wall and the hepatic area when examining the radiogram of the abdomen of a sick infant, and upon diagnosis of such, vigorous antibiotics, supportive therapy and surgical intervention when indicated may result in more survivors.

Summary

Gas in the portal veins and the intestinal wall of infants can be detected on plain radiograms of the abdomen. Whatever the underlying basic disease may be, this sign indicates that bowel wall is seriously compromised. Contrary to earlier experience, these infants may survive if the diseased bowel section can be resected.

Literature concerning the previously reported forty-three pediatric cases is reviewed and nine new cases are added.

REFERENCES

1. Wolfe, J. N., and Evans, W. A.: Gas in the portal veins of the liver in infants: A roentgenographic demonstration with post-mortem anatomical correlation, *Am. J. Roentgen.* 74: 486, 1955.
2. Fred, L. H., Mayhall, C. G., and Harle, T. S.: Hepatic portal venous gas. A review and report on six new cases, *Am. J. Med.* 44: 557, 1968.
3. Schoor, S.: Small intestinal intramural air, *Radiology* 81: 285, 1963.
4. Arnon, R. G., and Fishbein, J. F.: Portal venous gas in the pediatric age group, *J. Pediat.* 79: 255, 1971.
5. Frates, R. E.: Incompetence of the sphincter of oddi in the newborn, *Radiology* 83: 875, 1965.
6. Sisk, P. B.: Gas in portal venous system, *Radiology* 77: 103, 1961.
7. Susman, N., and Senturia, H. R.: Gas embolization of the portal venous system, *Am. J. Roentgen.* 83: 847, 1960.
8. Miskin, M., and Reilly, B. J.: Gas in the intestinal wall and portal venous system in infants, *Canad. Med. Ass. J.* 101: 129, 1969.
9. Bilger, M.: Pneumatosis cystoides intestinalis in children, *J. Pediat.* 49: 445, 1956.
10. Berdon, W. E., Grossman, H., Baker, D. H., Mizrahi, A., Barlow, O., and Blanc, W. A.: Necrotizing enterocolitis in premature infant, *Radiology* 83: 879, 1964.
11. Gazin, A. I., Brooke W. S., Lerner, H. H. and Prize, P. B.: Pneumatosis intestinalis: Roentgen diagnosis and surgical management, *Am. J. Surg.* 77: 563, 1949.
12. Pagan-Carlo, J., and DeMonk, E. H.: Hepatoportal pneumatosis with mesenteric venous thrombosis in an infant. Report of a case, *Amer. J. Roentgen.* 91: 699, 1964.

13. Goldstein, W. B., Cusmando, J. V., Gallagher, J. J., and Hemley, S.: Portal vein gas. A case report with survival, *Amer. J. Roentgen.* 97: 220, 1966.
14. Verby, H. D., Castellino, R. A., Priedland, G. W., and Northway, W. H.: Portal vein gas complicating Hirschsprung's disease with enterocolitis, *J. Pediat.* 73: 599, 1968.
15. McCandless, R. G.: Portal vein gas: Grave prognostic sign, *Am. J. Roentgenol.* 92: 1162, 1964.
16. Barrett, A. F.: Gas in portal vein diagnostic value in intestinal gangrene, *Clin. Radiol.* 13: 92, 1962.
17. Stevenson, J. K., Graham, B. C., Oliver, T. K., and Goldberg, V. E.: Neonatal necrotizing enterocolitis. A report of twenty-one cases with fourteen survivors, *Amer. J. Surg.* 118: 260, 1969.
18. Shaw, A., Cooperman, A., and Fusco, J.: Gas embolism produced by hydrogen peroxide, *New Eng. J. Med.* 277: 238, 1967.
19. Stiennon, O. A.: Pneumatosis intestinalis in the newborn, *Am. J. Dis. Child.* 81: 651, 1951.
20. Koss, L. G.: Abdominal Gas Cysts (Pneumatosis Cystoides Intestinorum Hominis): Analysis with Report of Case and Critical Review of Literature, *Arch. Path.* 53: 523, 1952.
21. Wiot, J. F., and Felson, B.: Gas in the portal venous system, *Amer. J. Roentgen.* 86: 920, 1961.