

hydrolyzing lactose in adult life are carrying a mutation derived from natural selection. In regions where malabsorption in adults is prevalent, lactose-free milk should be given to children so as to avoid diarrhea related to lactose intolerance.

A metabolic and anatomic study of a case of Noonan's syndrome with intestinal lymphangiectasia. H. LAWRENCE VALLET, PHILIP G. HOLTZAPPLE, WALTER R. EBERLEIN, WILLIAM C. YAKOVAC, THOMAS MOSHANG, JR., and ALFRED M. BONGIOVANNI. *Univ. of Pennsylvania Sch. of Med., The Children's Hosp., Philadelphia, Pa.*

Major cardiovascular anomalies and cutaneous lymphatic defects have been described in both Noonan's and Turner's syndrome.

A 6½ year old male presented with anasarca, chylous pleural effusions, cutaneous lymphatic leakage, ascites and hypoproteinemia. Studies revealed findings consistent with the diagnosis of intestinal lymphangiectasia and a major thoracic lymphatic vessel leak. Investigations included X-ray studies of small bowel, Cr⁵¹-chloride turnover, stool fat analysis, small bowel biopsy, and effusion studies for electrolytes, cells, lipoprotein electrophoresis and chylomicron analysis. Low lymphocyte counts and immunoglobulins were also found.

Medium chain triglycerides and a low fat diet corrected the protein loss but a seemingly mild pre-existing cardiac lesion worsened, and dictated the need for corrective pulmonary valve surgery. He died following this procedure.

Post mortem studies revealed severe defects in most of the mesenchymal components of cardiovascular organogenesis. Large lympho-venous shunts were present in the lung, liver and pancreas.

The lymphatic defects in these patients may not be as benign as once believed, and may be a major cause of failure to thrive.

Transmissible gastroenteritis in piglets (TGE). A model for study of acute viral diarrhea. MARY KELLY, DANIEL G. BUTLER, and J. RICHARD HAMILTON, *Research Inst., Hosp. for Sick Children, Univ. of Toronto, Toronto, Ont., Can.*

We studied a specific viral gastroenteritis (TGE) in piglets in order to explore the pathogenesis of acute infectious diarrhea. We compared 23-26 day old piglets infected orally with TGE virus, with pair fed non-infected litter mates. Infected pigs lost weight; fecal weight, Na⁺, K⁺ and Cl⁻ excretion increased significantly; fat excretion did not increase. Serum concentrations of Na⁺, K⁺ and Cl⁻, Mg⁺⁺ and Ca⁺⁺ did not differ between groups. After 40 hours the pigs were killed. Although a mucosal lesion characterized by diffuse villous and epithelial cell damage occurred in some infected pigs, the groups did not differ significantly with respect to actual villous dimensions. In infected pigs the following changes in specific enzyme activity occurred: Na⁺-K⁺-ATPase and Mg⁺⁺-ATPase decreased significantly in proximal jejunum only; alkaline phosphatase decreased in proximal and mid-jejunum; sucrase decreased in mid-jejunum and ileum. There was no change in activity of any of these enzymes in proximal or distal colon. Mucosal protein content was the same in both groups. Our results suggest a relationship between proximal intestinal Na⁺-K⁺-ATPase activity and the diarrhea of acute viral enteritis.

Group	Stool Wt. (g/24 hr)	Stool Na (mEq/24 hr)	Enzyme activity—proximal jejunal mucosa (mean units/g. protein)			
	M. S.E.	M. S.E.	Na ⁺ -K ⁺ -ATPase	Mg ⁺⁺ -ATPase	Alk. Phos.	Su- crase
Control	36.3 ± 9.4	0.9 ± 0.6	1.35 × 10 ³	1.09 × 10 ³	8.43 × 10 ³	0.74
Infected	261.5 ± 74.4	22.0 ± 6.1	0.92 × 10 ³	0.74 × 10 ³	3.61 × 10 ³	0.38
P	<.05	<.01	<.01	<.05	<.01	>.05

Hereditary pancreatitis (HP) without amino-aciduria: Two new kindred. ALLEN LAPEY, JOHN KATTWINKEL, PAUL A. DI SANT'AGNESE, and LEONARD LASTER. *NIH, Bethesda, Md.*

HP, an autosomal dominant disorder with incomplete penetrance has been reported in 13 families and is characterized by chronic relapsing pancreatitis leading to pancreatic insufficiency, pancreatic calcifications, and at times diabetes. In 3 of the original families lysine-cystine amino-aciduria was present in some members regardless of pancreatic involvement.

Our West Va. kindred (total 55 members) had 8 definite and 12 suspected cases of pancreatitis, mean age of onset 5 yrs.: 7 of 8 definite cases had pancreatic lithiasis, in 3 instances before 13 yrs. of age. Our Tenn. kindred (total 110 members) had 9 definite and 12 suspect cases, mean age of onset 14 years, with 6 of 9 definite cases presenting calcifications. There was striking variation in age of onset from 18 mos. to 35 yrs. Steatorrhea and pancreatic deficiency tended to be a late complication but they were found as early as 20 yrs. of age. There was no good clue as to what brought on acute attacks.

In both kindred fecal fat, pancreatic enzymes, and serum amylase and lipase were assessed. Serum lipids and parathyroid function by calcium infusion were normal. All urinary amino acids were determined in 7 patients and 9 relatives and were normal in all instances.

HP is a generally unrecognized cause of pancreatitis in childhood and important in the differential diagnosis of recurrent abdominal pain and pancreatic lithiasis (e.g., from cystic fibrosis). It is speculated that two different genetic types of HP exist with and without amino-aciduria.

Recurrent pleural effusion: A complication of pancreatitis in childhood. FREDERIC B. KOPEL, IRWIN GRIBETZ and HAROLD GROTSKY. (Intr. by Alex J. Steigman). *The Mount Sinai Sch. of Med., New York, N. Y.*

While pleural effusion as a complication of pancreatitis has been described in adults, this entity has not been noted, in the English literature, in children. We have recently uncovered chronic pancreatitis as the cause of recurrent pleural effusions in an 8-year-old Puerto Rican male whose presenting complaint at another hospital was recurrent substernal and epigastric pain radiating to the left shoulder. Exhaustive investigation, including cultures of the pleural fluid, skin tests for typical and atypical mycobacteria and fungi, lupus preparations, bronchography and thoracotomy with pleural biopsy, failed to reveal the cause of the recurrent pleural effusions. Substernal pain recurred, and the initial complaint of epigastric pain was only then appreciated. Pleural fluid showed an amylase concentration of more than 1000 Somogyi units/100 ml at a time when the serum amylase content was 335 units/100 ml (normal = 30-180 units). Pancreatic stimulation with secretin (Boots) 1 unit/kg resulted in a 1 hour output of 25 ml or 1.4 ml/kg (normal = 2 ml/kg), and a maximal