2. Gilroy JA, Adams AB. Annular pancreas. Radiology, 1960, 75: 568-570.

3. Irving IM. Duodenal atresia and stenosis: Annular pancreas. *In:* Neonatal Surgery, 3rd end. Eds Lister J, Irving IM. London, Butterworths, 1990, pp 424-441.

4. Merrill JR, Raffensperger JG. Pediatric annular pancreas: Twenty years' experience. J Pediatr Surg 1976, 11: 921-925.

Alstrom Syndrome

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The Alstrom Syndrome is a rare autosomal recessive disorder with involvement of the retina, ear, kidney and endocrine glands. During childhood, Alstrom syndrome usually presents with blindness (due to retinal degeneration and pigmentary changes) and nerve deafness. In adults, carbohydrate intolerance and slowly progressive renal disease develops; while obesity may disappear(1,2). Males have hypogonadism with small testes and low plasma testosterone, but normal secondary sexual characteristics(3). Females lack evidence of hypogonadism but their menstrual periods are irregular. Other manifestations include hyperuricemia, hypertriglyceridemia(3), acanthosis nigricans(4) and baldness. This syndrome has never been diagnosed in childhood and only about ten cases in adults have been reported till date.

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Case Reports

Two siblings, born to Indian Muslim first cousin consanguinous parents, are being reported. Three other siblings and other members of the family were normal.

Case 1: A 13-year-old girl was referred for evaluation of blindness and diabetes mellitus. She was delivered normally at term with an uneventful perinatal period. Parents suspected poor vision at 5-6 months of age. Till about 5 years of age she was able to recognize toys but subsequently visual acuity worsened and she became completely blind by 6 years. Parents noticed that she was obese at 6 years when compared to other children of her age. She also had hearing problems. One year back she attained menarche, but her periods were irregular and varied from scanty to hypermenorrhea.

On physical examination, the patient was a short statured, mildly obese, intelligent girl. Her height and weight were 140 cm (3rd centile) and 41 kg (25th centile), respectively. Scalp and axillary hair were sparse. There was no evidence of baldness. Pubic hair and breast development were normal for her age (SMR Stage 3). She was completely blind and deaf. Extremities were normal, with no digital anomalies.

Opthalmological examination revealed bilateral dilated and fixed pupils with horizontal nystagmus. Optic fundii showed

bilateral generalized retinal atrophy with clumps of brownish pigment deposition in the vascular area.

Audiometry revealed moderately severe sensorineural hearing, impairment of higher frequencies. No anatomical abnormality of the ears was observed.

On investigations urine showed a sugar more than 1 g/dl, albumin and acetone being absent. Urinary specific gravity and pH were 1.020 and 6.0, respectively. Routine hematology, liver functions, blood urea, serum creatinine, serum uric acid, *X*-ray chest, EEG, echocardiogram and CT scan of the brain were normal. Fasting blood sugar was 12.71 mg/dl while a two hour post prandial blood sugar was 16.04 mg/dl. Serum triglyceride level was 3.9 g/L.

Case 2: This 10-year-old male was the younger brother of Case 1. He was delivered full term of an uneventful pregnancy. His poor vision was noticed at about 6 months of age. This worsened and at the time of admission even finger counting beyond one foot was not possible. According to the parents he was very obese in his early childhood.

The patient was a short statured intelligent boy with articulate speech. His weight and height were 20 kg (25th centile) and 124.5 (3rd centile), respectively. Scalp hair were normal. His penis and testes were small for his age (SMR Stage 1). Extremities were normal and he had no digital anomalies. Systemic examination revealed blindness and sensorineural deafness. There was no anatomical abnormality of the ears.

Ophthalmological evaluation revealed that he could count fingers at 25 cm. Pupils were semidilated and reacting to light. Fundoscopy revealed a pale waxy disc, marked mottling and attenuation of vessels and macular degeneration. No pigment deposition was seen. Audiogram revealed marked sensorineural hearing loss in all frequencies.

Routine hematology, urinalysis, fasting blood sugar, glucose tolerance test, *X*-ray chest, blood urea and serum creatinine were normal.

Discussion

As in all previously reported cases(1-4) both our cases had blindness, deafness and infantile obesity. The elder one also had carbohydrate intolerance which is a feature of the Alstrom Syndrome but generally appears in the second or third decade.

Intelligence was normal in both our cases which is consistent with the previously reported cases. In the first case, a female, secondary sexual characters were normal for her age, while the boy had testes which were small for his age. This is also in accordance with the previously reported cases(3). Chronic nephropathy was not recognized in Alstrom's initial report, but later this was reported. However, this was a feature which developed in adulthood(2). Our cases showed no evidence of nephropathy. It is possible that they may develop this feature as their ages progress.

Hypertriglyceridemia(2) was present in our first case. This has been reported in previous cases. Two of the five sibships reported previously resulted from wedlock between distant cousins(1,4). Parents of our siblings are first cousins.

Although, superficially, the Alstrom Syndrome may bear resemblance to the Laurence-Moon-Biedl Syndrome(4), absence of mental retardation and digital anomalies clearly separates the former from the latter. Moreover, the combined occurrence of nerve deafness, diabetes

mellitus and nephropathy is rare in Laurence-Moon Biedl Syndrome.

REFERENCES

1. Alstrom CH, Hallgren B, Nilsson LB, Asander H. Retinal degeneration combined with obesity, diabetes mellitus, and neurogenous deafness: A specific syndrome (not hitherto described) distinct from the Laurence-Moon-Bardet-Biedl Syndrome. Acta Psychiat Neurol Scand 1959, 34 (Suppl 129): 1-8.
2. Goldstein JL, Fialkow PJ. The Alstrom Syndrome. Medicine 1973 52: 53-55.
3. Weinstein RL, Kliman B, Seully RE. Familial syndrome of primary testicular insufficiency with normal virilization, blindness, deafness, and metabolic abnormalities. New Eng J Med 1969, 281: 969-972.
4. Klein D, Ammann F. The syndrome of Laurence-Moon-Bardet-Biedl and allied diseases in Switzerland: Clinical, genetic and epidemiological studies. J Neurol Sci 1969, 9: 479-483.

Cholecystitis and Cholelithiasis in Children

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Cholecystitis and cholelithiasis are uncommon in children. Although the Western literature appears to suggest an increasing incidence of cholelithiasis in children, Indian studies are not available to substantiate this. In an Indian study of 228 cases of cholelithiasis and cholecystitis spread over a 15-year period the authors came across only one case aged 13 years(1). More recently in another study of 55 cases involving evaluation of cholecystitis both calculus and acalculus, in young patients, the authors reported only two cases in the age group of 0-10 years(2). It is, therefore, not surprising that biliary tract is often overlooked as a possible cause for abdominal pain in children(3). However, even with the increased awareness of cholecystitis and cholelithiasis in the pediatric age group there is still a lack of clear understanding of the etiology, natural history and a standard treatment in these patients. We report 3 cases of cholecystitis and cholelithiasis encountered over a 1½ year period in the age group of 4-12 years.

Case Reports

Case 1: A 4-year-old female child presented with recurrent attacks of right upper quadrant abdominal pain associated with vomiting of 4 months duration and she was afebrile during the attacks of pain. General and systemic examination was within normal limits. On abdominal examination, a soft, smooth, hepatomegaly, 4 cm in the midclavicular line, was the only positive finding. On investigations, the hemoglobin, the total and differential leucocyte counts and the liver function tests were normal. Peripheral blood smear was within normal

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