FIG.—Progress of the patient before and during treatment with DDAVP. ▲ urine osmolality, ○ plasma osmolality. Stippled area is range of urinary specific gravities. Open columns show daily fluid intake, solid columns show daily urinary output.

The drug was withheld for several days and the patient's symptoms recurred. Treatment was started again on day 8 (point C in Fig.) and was thereafter continued. She was able to concentrate her urine satisfactorily on a schedule of 5μg DDAVP twice daily. No side-effects were noted, and the patient easily administered the preparation with confidence. The patient has since remained well.

Discussion

DDAVP is a new synthetic analogue of vasopressin administered intranasally via a plastic nasal catheter ('rhinyle'). A measured quantity of a solution of the preparation is collected in the 'rhinyle', one end of which is inserted through the anterior nares; the other end is placed between the lips, and the patient then blows the contents of the 'rhinyle' onto the nasal mucosa, from where the substance is absorbed.

In view of its ease of administration, freedom from side effects, and relatively long duration of action, DDAVP seems to be a useful preparation in the treatment of pituitary diabetes insipidus in children.

Summary

Idiopathic pituitary diabetes insipidus in a 10-year-old girl was successfully treated with a new synthetic analogue of vasopressin, 1-deamino-8-d-arginine vasopressin, given intranasally.

I am grateful to Dr. G. M. Komrower for permitting me to present his patient, and to Ferring Pharmaceuticals, Sweden, who supplied the DDAVP.

REFERENCE


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Oesophageal atresia

3 cases in 2 generations

We report what we believe to be the second recorded case of oesophageal atresia in parent and child. Engel et al. (1970) described a woman, one of their first successfully-treated patients, whose daughter was also affected. Both had oesophageal atresia with a tracheo-oesophageal fistula to the lower oesophageal pouch. This combination of anomalies is generally known as oesophageal atresia type III, and accounts for 85 to 90% of patients (Freeman, 1969).
Case reports

The pedigree is shown in the Fig.

Fig.—Pedigree of affected family. Symbol with / indicates a death.

Case II.3. Born 18 May 1947 after 36 weeks' gestation, weighing 1-7 kg. Labour had been induced early because of hydramnios. The child regurgitated all her feeds and oesophageal atresia was diagnosed. Right thoracotomy was performed (Professor R. S. Pilcher) on 21 May. There was no tracheo-oesophageal fistula. The ends of the oesophagus were too far apart to anastomose directly, therefore the stomach was mobilized and brought up into the chest in order to perform oesophago-gastrostomy. Copious clear fluid was found in the stomach and a presumptive diagnosis of duodenal atresia was made. The child's condition precluded further surgery and she died 2 hours after operation. At necropsy duodenal atresia proximal to the biliary sphincter was confirmed, other abnormal findings being an anteriorly-displaced anus and malrotation of the midgut.

Case II.4. Born 22 September 1948 at term weighing 3·8 kg after an apparently normal pregnancy without hydramnios. She regurgitated all her feeds and oesophageal atresia was diagnosed. Right thoracotomy was performed (Professor R. S. Pilcher) on 24 September. The ends of the oesophageal pouches were found to be overlapping. There was a fistula to the lower pouch and this was divided and the trachea repaired. It was possible to perform a tension-free anastomosis. The child was given intravenous fluids for 5 days and by the tenth day was taking normal feeds orally. She has remained trouble free to this day.

Case III.1. Born 24 September 1972, weighing 2·64 kg after a normal pregnancy without hydramnios, and normal delivery. In view of the family history an attempt was made to pass a nasogastric tube, though at this time oesophageal atresia was not suspected on clinical grounds. It was thus that the diagnosis was made.

Right thoracotomy was performed on 26 September when it was found that he had the common type of atresia with fistula to the lower pouch. The fistula was divided and it was possible to perform a primary anastomosis, but with a little tension on the oesophageal suture line.

He initially did well but the anastomosis strictured and excision of the stricture and re-anastomosis was performed one month after the first operation. His swallowing was satisfactory after this but he developed an incisional hernia of the chest wound which was repaired at 5 months of age. His subsequent course has been satisfactory.

Thus Case III.1. and his mother had oesophageal atresia type III without associated malformations, while the mother's sister, Case II.3, had oesophageal atresia without fistula (type I), duodenal atresia, ectopic anus, and malrotation of the midgut.

Discussion

The population incidence of oesophageal atresia is about 1 in 3000 livebirths (Freeman, 1969). There appear to have been no systematic studies of familial incidence, but there are 4 reports of 2 affected sibs (Grieve and McDermott, 1939; Lamman, 1940; Copleman, Cannata, and London, 1950; Haight, 1957). Sloan and Haight (1956) were told of 2 further sib pairs, one of which consists of our Cases II.3 and II.4. There are 2 accounts of families in which 3 sibs had oesophageal atresia; in the first (Hausmann, Close, and Williams, 1957), 2 children with the type III anomaly were followed by 1 with type I, plus cleft lip and palate. In the second (Forrester and Cohen, 1970) all 3 children had type III oesophageal atresia, all had an absent or ectopic anus, and at least 2 had a laryngeal fissure. There is no mention of parental consanguinity, but in view of its consistency within the family it is possible that this represents a distinct entity.

Ten of Haight's cases were twins, 6 were thought to be identical, and in each case the other twin was unaffected. Haight found a further 6 pairs of twins in published reports, and all were discordant for the anomaly. There are, however, later reports of 2 pairs of identical twins where both were affected (Woolley, Chinnock, and Paul, 1961; Blank, Prillaman, and Minor, 1967). In each case both twins had the type III anomaly.

It is clear that oesophageal atresia is not determined, in the majority of cases, by an allele or alleles at a single locus. Environmental and polygenic factors probably contribute to liability to develop the anomaly, as has been postulated by Carter (1969) for cleft lip, with or without cleft palate, and pyloric stenosis. It is known that such a mechanism will simulate single gene inheritance in occasional families possessing many of the appropriate alleles. The high frequency of associated malformations, and the occurrence within our family and that of Hausmann et al. (1957) of 2 different varieties of atresia, may indicate that the genetic contribution is relatively nonspecific.
Short reports

With the first successfully treated patients in this country now in their mid-20s, it should soon be possible to obtain a direct estimate of the risk to the children of affected individuals. It seems reasonable to advocate in the children of such affected parents that the continuity of the oesophagus should be confirmed by passing a nasogastric tube before starting feeds.

Summary

A family is reported in which a boy, his mother, and his mother’s sister all had oesophageal atresia. Previous reports of familial occurrence of the condition are reviewed, and it is concluded that the aetiology is, in most cases, multifactorial.

We thank Mr. D. J. Waterston for permission to report his patient.

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New method for arterial blood sampling in infants and adults

The conventional methods for obtaining arterial blood samples for gas analysis are not easy, nor are they free from certain potential errors. It is difficult to fill the dead space of a syringe with heparin solution without including small air bubbles which may also tend to stick between the barrel and the plunger. Finally, in newborn infants more blood tends to be withdrawn than is needed for the actual measurements. For these reasons, a convenient method has been devised for obtaining the small volume of arterial blood which is often all that is required for gas analysis.

Material and methods

A number 17 needle is connected by means of a short piece of silicone tube to a dry heparinized glass capillary tube having an outer diameter of 1.4 mm, a length of 100 mm, and a capacity of 50 µl (Fig.).

FIG.—Device for puncture.

Depending upon the amount of blood needed, 1, 2, 3, 4, or even 5 capillaries are connected with a silicone tube. If large volumes are required there are capillaries each having a capacity of 150 µl.

With this system we have performed punctures of the radial artery both in newborn infants and in adults. The radial artery lies on the radial side of the tendon of flexor carpi radialis, where in the infant repeated attempts may

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