INFANTILE PYLORIC STENOSIS

AN ESSAY

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By

AMR MOHAMED ABOUL - FADLE

M. B., B. Ch.

Supervised by

Prof. Dr. AHMED FAWZY BAHNASSY

Prof. of General Surgery

Dr. IMAM FAKHR

Lecturer of General Surgery

Faculty of Medicine

Ain Shams University

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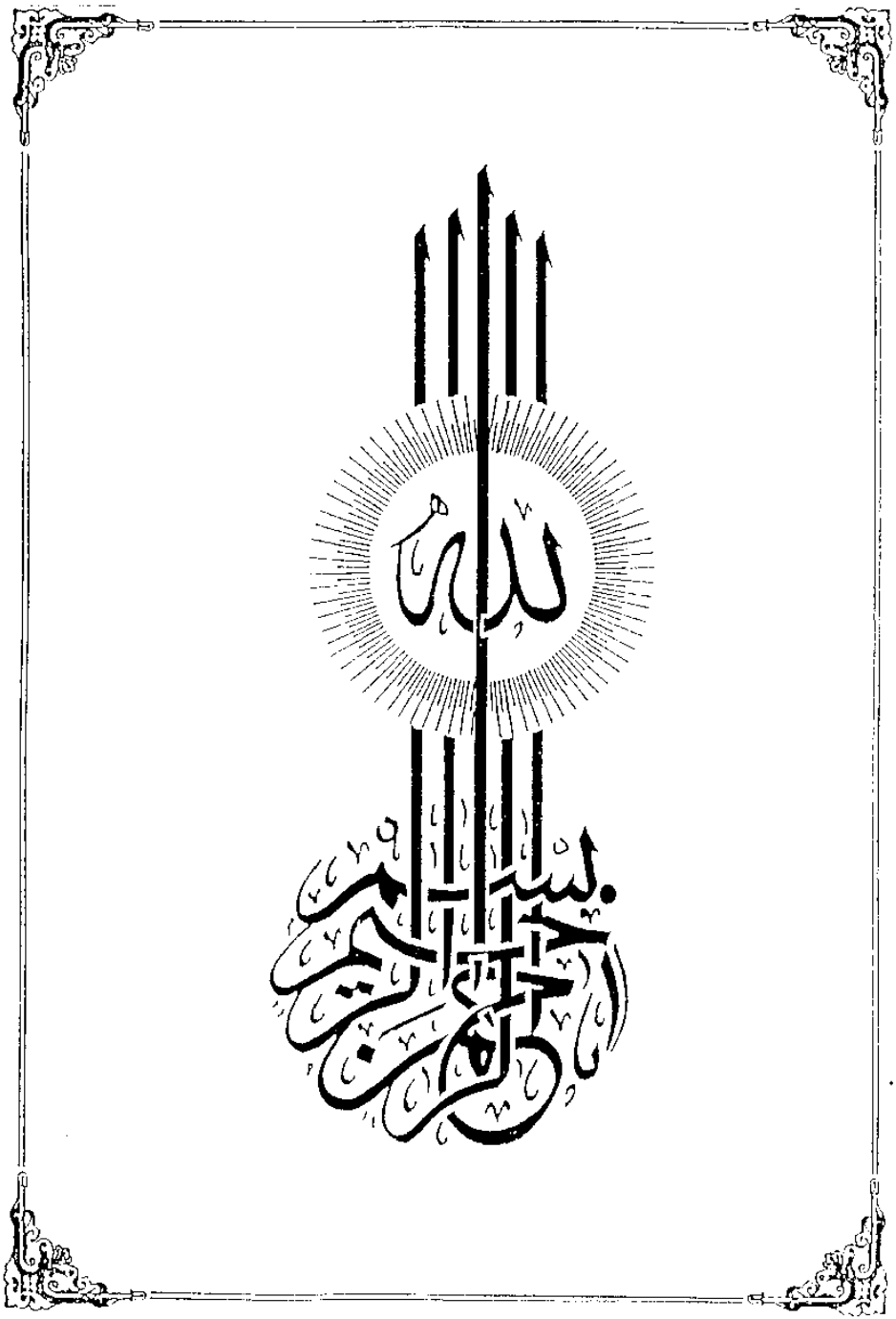

C O N T E N T S

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I N T R O D U C T I O N

Infantile pyloric stenosis is a relatively common problem in the early infancy. It is one of the urgent intraabdominal conditions requiring surgery in the first few weeks of life. Vomiting, at about the third week of life, is the presenting symptom. The Ramstedt operation is the treatment of choice with minimal morbidity and zero mortality. Many theories have been made to know the aetiology of the disease but, it is still obscure.

HISTORY AND INCIDENCE

H I S T O R Y

The first description of the characteristic clinical and post-mortem findings of congenital hypertrophic pyloric stenosis was by Patrick Blair, a Scots surgeon and botanist, who presented his case to the Royal Society in London in 1717 (Blair, 1717).

World wide interest in the disease was not aroused until 1888 when Harald Hirschsprung, the Copenhagen Paediatrician, described the clinical and post-mortem findings in two children (Hirschsprung, 1888).

Following Hirschsprung's description of congenital hypertrophic pyloric stenosis, an interesting number of cases were reported, a total of 598 up to 1910 and, whilst Hirschsprung himself made no suggestions concerning treatment, from this moment onwards, attempts at treatment began, initially medical but very soon surgical as well. Medical treatment with thickened feeds and atropine reduced the mortality slightly and was continued well into the early twentieth century since the results of the early surgical treatments were little better. (Spicer, 1982).

Surgical treatment of pyloric stenosis in those early days was of three kinds, gastroenterostomy, dilatation of the pylorus and various forms of pyloroplasty. (Mack, 1942).

The first successful operation for pyloric stenosis was a gastroenterostomy performed in 1898 by Lobkar, and other authors reported cases treated by this operation over the next 10 years, but with a considerable mortality (Spicer, 1982).

In 1899, Nicoll of Glasgow successfully treated a case by dilatation of the pylorus via a gastrotomy but other surgeons who tried this method found it too hazardous. (Spicer, 1982).

Pyloroplasty was first performed in 1903 by Dent of London. (Dent, 1904).

Extramucous pyloroplasty was developed, apparently independently, by Nicoll of Glasgow in 1906, Fredet of Paris in 1907 and Weber of Dresden in 1908. This operation was undoubtedly an advance, but all these surgeons felt it essential to suture the divided muscle transversely. It was against this background that in

1911 Ramstedt first performed the operation which bears his name and is still the standard treatment for infantile pyloric stenosis.

Ramstedt discovered the operation almost accidentally (Pollock and Norris, 1957). He reported it the following year and published these two successful cases. (Ramstedt, 1912).

Within 10 years the operation gained wide acceptance with published mortalities below 10 percent. The mortality has declined steadily ever since; in a more recent series of 1465 cases treated over a 29 years period, the mortality was 0.4% (Benson, 1970).

The prognosis of babies with pyloric stenosis has been improved by the improved general health of all infants, better anaesthesia and particularly by the more precise management of fluid imbalance (Spicer, 1982).

I N C I D E N C E

Infantile pyloric stenosis occurs in 2.6 of 1000 Live births. (Dodge, 1975).

The mean incidence of infantile pyloric stenosis in the United Kingdom has been approximately 2.5 per thousand live births for many years. However, there is recent evidence that the incidence is increasing. The incidence in the Trent region in 1978 was 2.5/1000 but rose to 3.8/1000 in 1979, and figures for 1980 suggest that this higher rate is being maintained (Spicer, 1982).

A recent publication from Scotland, showed an even more marked rise, 5.2/1000 in 1978 and 8.8/1000 in 1979, the latter figure being the highest ever recorded in the world. (Kerr, 1980).

Geographical variations in incidence are not great between European countries. The disease is slightly less common in North America and is relatively rare in Negroes and babies of Chinese extraction (Klein and Cremin, 1970).

It is rare in India, an incidence of 1/3500 has been quoted as representative. (Joseph and Nair, 1974).

The average age of onset of symptoms is 3.5 weeks and most cases occur around this age, though more rarely symptoms may start at birth or be delayed until 3 months of age. The natural history is remission over a period of 2-5 months and the disease is not a significant problem after the first few months of life. (Spicer, 1982).

Several cases have been reported with onset of symptoms and eventual operation within the first day or two of life, and indeed one case described, with vomiting since birth and preceding polyhydramnios may have been a case of pyloric stenosis in utero. (Powell, 1962).

Certainly, the typical post-mortem findings have been described in neonates, (Meeker, et al., 1948) and even in the fetus. (Dent, 1907; Green and Sidbury 1919).

At the other end of the age scale, cases have been reported at the age of 3.5 years, 6 years and 12 years. (Couvreur and Gerbeaux, 1968). These late childhood cases merge into the entity of adult hypertrophic pyloric stenosis, a disease of great interest in its own right. (Du Plessis, 1966; Keynes, 1965). If one considers true primary adult hypertrophic pyloric stenosis (and not those cases which are secondary to peptic ulceration) the onset of symptoms is often insidious and sometimes may be traced back to childhood.

Infantile pyrloric stenosis is more common in boys than in girls . (Spicer, 1982).

Peaks of incidence in the spring (April to June) and autumn (September to November) are apparent in many of the published series (Spicer, 1982).

Mothers of affected children have been found to be significantly younger than mothers of control children and there is general agreement that breast-fed babies are more prone to the condition than bottle-fed babies, though whether this is direct cause

and effect has not been established. (Swiatkowska, 1977).

A decreased incidence in subjects of blood group "A" is recorded. (Dodge, 1967).

An association with oesophageal atresia has been found. Vilarino and co-workers reported an incidence of 1 in 60. (Vilarino et al., 1977).

Hiatus hernia is associated much more commonly than would be expected by chance, a 13 percent incidence being quoted by Pellerin (Pellerin et al.,1974). Whether this is a primary association or whether it is a result of the prolonged vomiting is not clear.

As regards chromosomal abnormalities, a significantly increased incidence of pyloric stenosis is associated with ovarian dysgenesis (Benson et al., 1964) and x/xx mosaicism (Vanderhorst et al., 1971).

An association with dominantly inherited polycystic kidneys has been described (Loh et al., 1977),

malrotation and the Smith-Lemli-Optiz syndrome (Gellis and Feingold, 1968).

A retrospective study noted 5 cases of infantile hypertrophic pyloric stenosis following ingestion of erythromycin (San Fillipo, 1976).

There is no doubt that there is a strong hereditary factor in the etiology of pyloric stenosis. There are many instances of high familial incidence. (Terezis et al., 1959, Keizer, 1952 and Fenwick, 1953).

There is slightly increased risk of both twins being affected in the case of monozygotic twins and a lesser risk in the case of non-identical twins. (Spicer, 1982).

The definitive work on the inheritance of pyloric stenosis is that of Carter. He concluded that the mechanism of inheritance is polygenic (Carter, 1961, Carter and Evans 1969).

An increased incidence in the first born has been quoted for many years (Spicer, 1982).