INFANTILE HYPERTROPHIC PYLORIC STENOSIS – A RARITY IN MALAYSIA?

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SUMMARY

Sixty seven infants were admitted over a 10-year period from 1972 to 1981: 50 males and 17 females. A higher incidence of hypertrophic pyloric stenosis exists in West Malaysia than was previously recognised. However, it remains at less than one tenth of the Western figures. Among the three major races, the condition was commonest in Indians and least in Malays. Most cases presented between the ages of 3 to 8 weeks. Prolonged history of vomiting of more than 3 weeks occurred in 43.3 percent cases, usually among Malay and Indian patients. All patients were treated surgically with 1 death (1.5 percent). The usual postoperative complication was vomiting which occurred in 22.4 percent. Early presentation, adequate preoperative resuscitation and improved anaesthetic techniques can further reduce this mortality and morbidity.

INTRODUCTION

Infantile pyloric stenosis was probably first described by Hildanus in 1646. [1] Blair [2] was the first to publish autopsy findings but our present day understanding of the disease is largely due to the work of Hirschsprung. [3]

The condition has a strong genetic influence [4,5,6] and variation in racial susceptibility exists. It appears to be rare in Chinese [7] and Indians. [8] In West Malaysia, it was reported to be very uncommon. [9] This paper summarises our 10-year experience with 67 Malaysian infants with this condition.

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MATERIALS AND METHOD

Records of all infants admitted to the General Hospital Kuala Lumpur diagnosed as having pyloric stenosis and confirmed at operation from January 1972 to December 1981 were examined. There were 50 boys and 17 girls giving a male to female ratio of 3 : 1.

RESULTS

There was a significantly higher incidence among Indians than among the other two major races of the country. The lowest incidence was in Malays who constituted 19.4 percent of the cases (Table I). All patients were aged 4 months or under. Fifty-four (80.6 percent) presented between the ages of 3 to 8 weeks, with a peak at 4 weeks (Fig. 1).

Vomiting was a constant feature. In the majority

TABLE I
ETHNIC GROUPS AND HYPERTROPHIC PYLORIC STENOSIS

	Malay No.	%	Chinese No.	%	Indian No.	%	Total No.
With HPS	13	19.4	22	32.8	32	47.8	67
Total paediatric surgical admission	2548	35.4	2672	37.2	1973	27.4	7193
					$x^2 = 13.988$; $p < 0.01$		

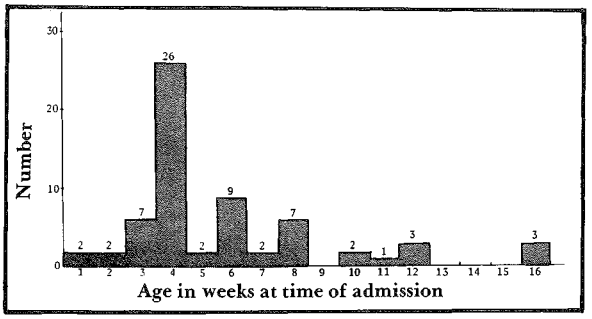

Fig. I Age at time of admission of 67 patients with HPS

in was projectile in nature and in none did the vomitus contain bile (Table II). Constipation and weight loss were presenting symptoms in almost half the cases. Epigastric peristalsis was observed in 43 patients (64.2 percent) and a pyloric mass was felt in 41 (61.2 percent). One 2-week old baby was jaundiced. Plain abdominal x-rays were done in 37 cases (Fig. 2) and in 12 cases barium meal was necessary to confirm the diagnosis (Fig. 3). Majority of the patients were admitted with history of vomiting for more than one week and in 29 (43.3 percent) it was for longer than three weeks (Table III). Delay in seeking treatment commonly occurred in Malay and Indian patients. There was no family history of the disease.

All patients had a standard Fredet-Ramstedt's pyloromyotomy performed under general anaesthesia, 10 via a paramedian, 31 via a midline and 26 via a transverse incision. Five accidental

TABLE II
CLINICAL FEATURES IN 67 PATIENTS WITH PYLORIC STENOSIS

Feature	No.	%
Symptoms		
Vomiting	67	100.0
Constipation	31	46.3
Weight loss	29	43.3
Epigastric distension/ abnormal movements	15	22.4
Physical Findings		
Epigastric peristalsis	43	64.2
Pyloric mass	41	61.2
Dehydration	34	50.7
Jaundice	1	1.5
No. of Abdo. x-rays done	37	55.2
No. of barium meals done	12	17.9

TABLE III
LENGTH OF HISTORY IN RELATION TO ETHNIC GROUPS

Length of History	Malay No.	%	Chinese No.	%	Indians No.	%	Total No.	%
<1 wk.	1	7.7	3	13.6	5	15.6	9	13.4
1-3 wk	2	15.4	13	59.0	10	31.3	25	37.3
>3 wk	8	61.5	6	27.4	15	46.9	29	43.3
Un-known	2	15.4	0	0	2	6.2	4	5.9
Total	13	(100.0)	22	(100.0)	32	(100.0)	67	(100.0)

Average: 2.9 weeks

perforations of the duodenal mucosa occurred, one of which was missed at the initial operation. All perforations were closed and there were no deaths associated with this. Oral feeding was commenced within 24 hours in 54 (80.6 percent). Only in seven cases (10.4 percent) was feeding withheld for more than 48 hours due to complications.

The most common postoperative complication was vomiting (22.4 percent) (Table IV). However, in none did it last for more than one week. One patient had to be reoperated due to incomplete

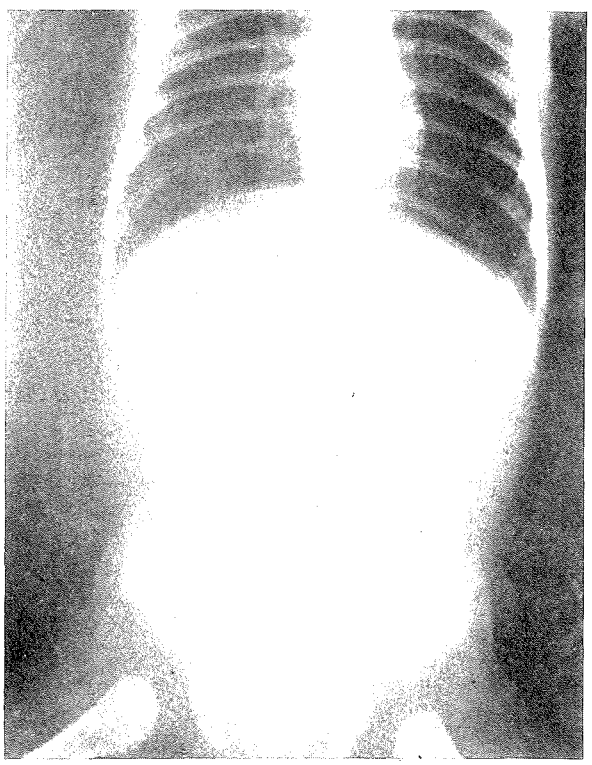

Fig. 2 Plain x-ray of abdomen showing distended stomach.

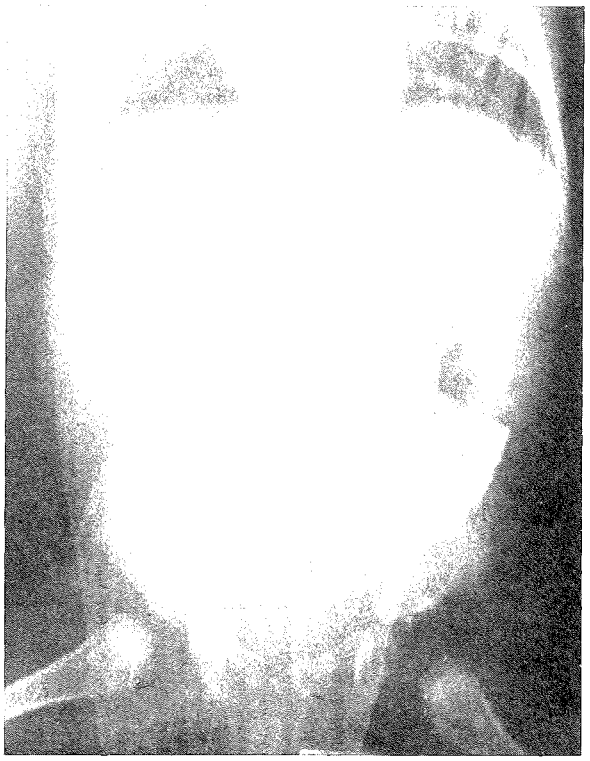

Fig. 3 The 'string sign' on barium meal.

myotomy. Another was reoperated because of a missed duodenal perforation. Both these babies survived. The one death was due to renal failure. Forty-seven patients (70.1 percent) were discharged home within a week of operation. Only 3 babies (4.5 percent) stayed between 2 to 3 weeks.

DISCUSSION

Hypertrophic pyloric stenosis (HPS) is the fourth most common cause of alimentary tract obstruction in infants under 1 year of age in this hospital. Between the ages of 1 to 3 months, it is the commonest obstructing lesion making up almost a quarter of the cases of obstruction seen in this age group (Laidin, to be published). Approximately 10 new cases are seen annually in Kuala Lumpur in the three hospitals treating paediatric surgical patients, namely the University Hospital, [10] the Pantai Medical Centre [4] and the General Hospital. Since these three hospitals cater mainly for the Selangor state which has approximately 60,000 live-births yearly, the incidence of the disease is estimated to be about 1 : 6000 livebirths. Chong and Lee [12] found an incidence of 1 : 5000 livebirths in Singapore. Field [9] estimated the incidence in

TABLE IV
POSTOPERATIVE COMPLICATIONS IN THE 67 OPERATED CASES OF PYLORIC STENOSIS

Complication	No.	%
Vomiting > 24 hrs.	15	22.4
Diarrhoea	4	6.0
Respiratory problems	2	3.0
Wound infection	1	1.5
Wound dehiscence	1	1.5
Bile peritonitis	1	1.5
Death	1	1.5
Total No. of complications	20	29.9
Total No. of patients with complications	18	26.9

West Malaysia to be 1 : 100,000 livebirths. This increase may not be entirely due to inadequate reporting in the earlier study. Murfin [13] found a true increase among Barbados Negroes over a period of 10 years. Incidence of the condition among Caucasians is approximately 1 : 400. [14,15,16] Therefore in this country, the incidence appears to be less than one-tenth of the Caucasian rate.

The incidence in India is approximately 1 : 3500. [17] In this study, almost half of the cases (47.8 percent) were Indians. Malay babies had the lowest incidence. Similar observations were made by Chong and Lee [12] in Singapore. This is in contradistinction to intussusception where the incidence is lowest among Indian and highest among Malay infants. [18] Besides genetic predisposition, environmental factors, e.g. culturally influenced feeding habits, may be important. Racial groups with low susceptibility who migrate to Western countries continue to have a low incidence of HPS. [7,19]

Many large series show a preponderance among first-born children. [5,7,15] This is especially so in male infants. [6,20] However, other reports have disputed this observation. [21,22,23] In this study, the size of the sample precludes any accurate conclusion to be drawn but no such predilection was apparent.

In this study, there was considerable delay in seeking treatment. Whereas less than 25 percent of patients in the West present with symptoms of more than 2 weeks, [24] almost 60 percent in this country do so. This is very similar to the Indian experience. [25] Besides weight loss and constipation, prolonged vomiting results in severe dehydration and electrolyte imbalance. A period of careful

assessment and prompt fluid and electrolyte correction is essential prior to operation which should be done soon but not necessarily as an emergency. [26] Our single death could be ascribed to inadequate preoperative resuscitation.

Projectile vomiting, visible 'golf ball' epigastric peristalsis and a palpable pyloric mass make the condition unmistakable. In most Western reports, the latter is detected in 80 to 95 percent of cases. In this series perhaps as a result of insufficient experience with the disease [24] the lump could be felt in only 61.2 percent (Table IV). It is sometimes necessary to give a test feed of 40 - 60 ml of water before epigastric peristalsis and the mass become apparent. A plain abdominal x-ray may be useful. A distended stomach with very little gas shadow elsewhere (Fig. 2) makes aerophagia, gastric volvulus and hiatus hernia unlikely. Barium meal and screening should only rarely be indicated. Majority of the barium studies were done prior to referral. The 'string sign' of Meuwissen and Slooff [27] is diagnostic (Fig. 3).

Jaundice due to unconjugated hyperbilirubinemia which subsided postoperatively was found in one patient. The association of jaundice with HPS was first pointed out by Martin and Siebenthal. [28] It occurs in about 2 percent of cases. [29] Arias *et al* [30] noted that the jaundice was usually due to unconjugated bilirubin and postulated reduced glucuronyl transferase activity as the likely cause. This has subsequently been confirmed. [29] Bleicher *et al* [31] suggested that elevated serum gastrin levels which is seen in some HPS babies [32] may affect glucuronyl transferase activity by some as yet unknown mechanism.

Non-surgical management of HPS is still being practised in parts of Sweden. It however involves more nursing hours and hospital stay of several weeks. This unnecessarily exposes the infant to cross-infection. [33] Follow up of surgically treated babies has not shown any serious long-term complications, [34] but there was a higher morbidity after medical management. Although peptic ulcers are not more common, gastritis and vague epigastric pain occur in 10-20 percent of cases, [35] especially within the first seven years. [33] The prolonged period of starvation entailed in this form of management may retard growth, produce permanent alteration in intelligence and personality development and may also result in subfertility. [36] Surgery has therefore been almost universally accepted as the preferred form of treatment for HPS. In this country especially where most of the babies come in a state of prolonged inanition, there is no place for medical management.

Although various incisions have been used, the preferred approach now is a right upper transverse, muscle-cutting incision which heals well with no complications. It gives good exposure and does not increase operating time or blood loss. Nine cases required blood transfusion due to pre-existing anaemia but this should not be necessary for the operation itself. Bleeding from the myotomy site will stop on returning the pylorus to the abdomen. [37]

Vomiting was the most common post-operative complication (Table IV). This is usually due to mucus gastritis which will subside with stomach washouts. Fifty-four infants (80.6 percent) tolerated feeds within 24 hours of operation. Incomplete myotomy in one infant resulted in prolonged vomiting. The reoperation done was probably unnecessary as many of these cases will improve after two or three weeks of conservative management. Duodenal mucosal perforation at the time of myotomy is not always fatal [15] and this is well illustrated in our five cases. It should however be detected and closed at the time of operation. A missed perforation can be lethal and should be suspected in a child who develops abdominal distension soon after operation.

Mortality of 1.5 percent is high by Western standards which is at present well below 1 percent. [26] Improvements in preoperative resuscitation and anaesthetic skills will undoubtedly allow these standards to be attained in the near future in this country.

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