Clinical Presentation of Pyloric Stenosis: the Change is in Our Hands

Ron Shaoul MD[1]*, Benjamin Enav MD[1]*, Zvi Steiner MD[2], Jorge Mogilner MD[2] and Michael Jaffe MD MRCP[1]

Departments of [1]Pediatrics and [2]Pediatric Surgery, Bnai Zion Medical Center, Haifa, Israel
Affiliated to Technion Faculty of Medicine, Haifa, Israel

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Abstract

Background: Hypertrophic pyloric stenosis classically presents as projectile vomiting during the third to fourth week of life, associated with good appetite. Additional classical presenting findings include palpation of the pyloric tumor, described as olive-shaped; a visible gastric peristaltic wave after feeding; and hypochloremic, hypokalemic metabolic alkalosis. It was recently claimed that this presentation has changed due to the easier access to gastrointestinal imaging.

Objective: To validate this contention and discuss possible reasons.

Methods: We conducted a retrospective chart review of all patients who underwent pyloromyotomy for HPS between 1990 and 2000. Only patients with confirmed HPS at the time of surgery were included. We also performed a comprehensive review of older studies for comparison.

Results: Seventy patients underwent pyloromyotomy over the 10 year period. Overall, 81% of patients were male infants and the mean age at diagnosis was 40 days. The mean duration of symptoms was 8 days. A firstborn child was noted in 43% of the cases. The classical symptom of projectile vomiting was absent in one-third of the patients, a pyloric tumor was not palpated in one-half of the cases, bicarbonate was higher than 28 mEq/L in 20%, and a pH of above 7.45 was present in 25% of patients. Hypochloremia was noted in about one-third. We found a good correlation between ultrasonographic width and length of the pylorus and the intraoperative findings. Pylorus length $\pm$ 24 mm correlated with significantly longer duration of symptoms. When compared with previous studies, the main findings were not significantly different; namely, mean age at diagnosis, percentage of male gender, and duration to diagnosis. The decrease in the number of pyloric tumors palpated paralleled the increase in the use of upper gastrointestinal series and ultrasonography in particular.

Conclusions: The clinical presentation of HPS has not actually changed despite the easier accessibility of GI imaging studies. However, the one significant change is the low percentage of pyloric tumors palpated, probably due to declining clinical skills accompanied by earlier utilization of imaging studies. The use of imaging and laboratory studies did not change the age at diagnosis but may have shortened the time for diagnosis and reduced the postoperative stay. Imaging and laboratory studies may be helpful for the subgroup with a non-classical clinical presentation.

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The incidence of hypertrophic pyloric stenosis is 1–8 per 1,000 live births and is more common in males by a ratio of 4:1 to 5:1 [1,2]. The etiology of pyloric stenosis is unknown. A localized lack of nitric oxide synthase, abnormal muscle innervation, and breast-feeding are possible factors leading to the development of HPS [1,3]. The classical presentation of HPS comprises an onset of regurgitation evolving to projectile vomiting during the third to fourth week of life associated with good appetite and constipation. Additional presenting findings classically include palpation of the pyloric tumor, usually described as olive-shaped, a visible gastric peristaltic wave after feeding, and hypochloremic hypokalemic metabolic alkalosis [1,4].

Since the advent of ultrasonography in 1977 by Teele and Smith [5] as a non-invasive efficient diagnostic modality for HPS, the confirmation of the diagnosis has become more efficient and is usually reached at an earlier stage of the evolving clinical picture [6,7]. Therefore, the well-described presentation of HPS has been claimed to change, at least in those areas where ultrasound examination or upper gastrointestinal imaging is available.

Although the first studies in ultrasonographic diagnosis of HPS appeared in 1977, the availability and accuracy of this method for diagnosing HPS has been customary since 1990. Over the years we have observed that many patients diagnosed with pyloric stenosis presented without the classical history, physical examination and laboratory studies, a presentation also suggested by others [6,8–10]. We undertook the present study to compare the changes in the clinical presentation of HPS in the 1990s in our institution with its readily accessible imaging studies to the previously described classical presentation of HPS and to evaluate any change and basis for change.

Patients and Methods

We conducted a retrospective chart review of all patients who underwent pyloromyotomy for HPS between 1990 and 2000, and included only patients with confirmed HPS at the time of surgery.

Data included demographic information, history of the illness, physical findings, admission laboratory values, and results of any imaging performed. The total and postoperative length of stay was calculated from the date of admission, surgery, and discharge. The data were analyzed using Microsoft Excel software. Statistical significance was determined by a P value < 0.05.

* Contributed equally to the preparation of this manuscript
HPS = hypertrophic pyloric stenosis

Results

During the 10 year study period 70 patients underwent pyloromyotomy for hypertrophic pyloric stenosis. Overall, 81% were male infants and the mean age at diagnosis was 40 days (range 10–193 days) with 56% presenting between 3 and 6 weeks of age. The mean duration of symptoms was 8 days (range 1–49 days). A firstborn child was noted in 43% of the cases (39% of boys and 61% of girls), and only 4% of the total patients had a family history of HPS. Prematurity (<37 weeks gestation) was present in 10% of patients. Seventeen percent of the babies were exclusively breast-fed, 45% were on cow's milk formula and 38% used both.

The classical symptom of projectile vomiting was absent in one-third of the patients; most of them had non-projectile vomiting or regurgitation for varying lengths of time. Furthermore, 63% of patients (17% overall) without projectile vomiting did not have a palpable pyloric tumor. No differences were noted between the groups (with and without projectile vomiting) with respect to age at presentation, length of symptoms and operative time. Although interval to operation was shorter in the projectile vomiting group (1.78 vs. 2.25 days) this was not statistically significant. Fever (4%) and diarrhea (7%) were uncommon in the clinical presentation.

The common physical finding of an olive-shaped pyloric tumor was not palpated in one-half of the cases. Its presence did not correlate with length of symptoms, and although its absence was associated with longer delay until surgery (2.42 vs. 1.48 days) this was not statistically significant. Clinical dehydration was present in 18% of the infants and did not correlate with length of symptoms, and only 4% had a serum urea level above 40 mg/dl.

Not surprisingly, as shown in Table 1, metabolic abnormalities at admission were found more commonly and were more noticeable in the absence of projectile vomiting. Of the patients who presented without projectile vomiting 58% had a chloride level below 98 mEq/L as compared to 32% of those with projectile vomiting. Unlike previous studies [11], bicarbonate was higher than 28 mEq/L in 20% of patients and pH was above 7.45 in only 25%.

The time from admission to the operating room was less than 24 hours for 59% of patients and 80% underwent pyloromyotomy within 48 hours. The mean total hospital stay was 6 days and mean postoperative stay 4.2 days.

Of the 70 infants, 69 had at least one imaging study, 42 had an ultrasound performed and 40 had undergone a barium study because sonography was not available. Sonography was introduced in our hospital in the late 1980s and the present series reflects the transition between both modalities. In eight cases a barium study was done after an ultrasound examination in order to confirm the diagnosis. As shown in Table 2, a very good correlation was seen for

Table 1. Comparison between patients with classical and non-classical presentations

Biochemical value	Projectile vomiting	No projectile vomiting	*P*	Olive palpated	No olive palpated	*P*
Cl- (mmol/L) Mean (SD)	98 (7.6)	96 (3.3)	0.021	98 (5.1)	97 (8.0)	0.437
HCO_3- (mmol/L) Mean (SD)	25 (4.3)	27 (5.3)	0.016	26 (5.2)	25 (4.3)	0.929
Cl- < 98 mmol/L	32%	58%		39%	40%	

Table 2. Comparison between ultrasound and intraoperative pyloric measurements

Measurement	Operative (mm)	Ultrasound (mm)
Pyloric thickness	4.07 ± 0.84	3.89 ± 0.20
Pyloric length	22.4 ± 2.82	21.4 ± 2.70

Table 3. Clinical parameters of HPS from other studies

Study	Mean age at diagnosis (days)	Mean length of illness (days)	Male (%)	Pre-mature (%)	Family history (%)	Olive pal-pate (%)	Postoperative hospital stay (days)
Benson [20]							
1940–62	21	NA	79	3	3	78	13.5 (1940–45)
(n=1,120)							3.9 (1961–62)
Cremin [21]							
1956–67	30	–	82	–	6	83	–
(n=165)							
Bell [18]							
1957–67	40.6	14.8	77	1.9	7.3	–	–
(n=305)							
Hulka [6]							
1969–74	41	12	85	–	–	79	5
1975–79	39	16	77	–	–	52	4
1980–84	35	7	83	4	12	43	3
1985–89	39	9	81	–	–	25	3
1990–94	38	8	81	–	–	23	3
(n=901)							
Papadakis [8]							
1973–75	38			–	–	–	4.4
1983–85	32	NA	81	–	–	–	3.4
1993–35	24			–	–	–	2.8
(n=283)							
Macdessi [10]							
1974–77	40	7	79	–	–	99	6.2
1988–91	40	8	82	–	–	79	3.1
(n=402)							
Breaux [9]							
1980–84	–	–	–	–	–	89	–
(n=216)							
Forman [17]							
1985–88	NA	NA	–	–	–	77	–
(n=101)							
Van der [16]							
Schouw 1990	39	11	70	–	13	–	–
(n=105)							
Smith [12]							
1991–92	34	11	–	–	–	73	–
(n=75)							
Hernanz-Schulman [19]							
1991–93	43	NA	–	15	–	–	–
(n=152)							
Chen [7]							
1993–95	30	6	–	–	13	–	–
(n=100)							
Shaoul							
1990–2000	40	8	81	10	4	50	4.2
(n=70)							

Table 4. Laboratory parameters of HPS in other studies

Study	Alkalosis (%)	Hypo-chloremia (%)	Imaging UGI or US (%)
Benson [20]			
1940–62	–	–	UGI 34%
(n=1,120)			
Cremin [21]			UGI 17%
1956–67	–	–	(only for non-palpable tumor)
(n=165)	–	–	
Bell [16]			
1957–67	–	–	UGI 84%
(n=305)			
Hulka [6]			
1969–74	–	–	UGI 55%
1975–79	–	–	UGI 90%
1980–84	–	–	UGI 90%, US 5%
1985–89	–	–	UGI 60%, US 35%
1990–94	–	–	UGI 40%, US 50%
(n=901)			
Papadakis [8]	HCO_3 > 28 mEq/L	CL < 90 mEq/L	
1973–75	12	12	UGI 27%
1983–85	15	14	UGI 67%
1993–95	10	9	US 96%
(n=283)			
Macdessi [10]			
1974–77	–	–	UGI 20%
1988–91	–	–	UGI or US 61%
(n=402)			
Touloukian [22]	HCO_3 > 30 mEq/L		
1977–81	24.6	–	–
(n=65)			
Breaux [9]			
1980–84	–	–	UGI 63%, US 26%
(n=216)			
Forman [17]			US 41%
1985–88			(patients w/ no palpable tumor)
(n=101)			
Smith [12]	$HCO_3 \geqslant$ 29 mEq/L	Cl $\leqslant$ 98 mEq/L	
1991–92	36	49	–
(n=75)			
Hernanz-Schulman [19]			
1991–93	–	–	US 100%
(n=152)			
Chen [7]	HCO_3 > 28 mEq/L	Cl < 90 mEq/L	
1993–95	9	10	US 96%, UGI 4%
(n=100)			
Shaoul	HCO_3 > 28 mEq/L	Cl < 98 mEq/L	
1990-2000	20	40	US 60%, UGI 57%
(n=70)		Cl < 90 mEq/L	
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ultrasonographic measurements of pylorus length and thickness and the operative findings. The length of the pylorus correlated with the duration of symptoms. A length $\geqslant$ 24 mm was associated with a significantly longer duration of symptoms (11.36 $\pm$ 11.07 vs. 4.4 $\pm$ 2.6 days).

Tables 3 and 4 summarize several studies over the past 50 years. We compared clinical and laboratory parameters such as age of diagnosis, length of illness, incidence in males and premature babies, family history, palpation of olive, postoperative stay, alkalosis, hypochloremia, and type of radiologic study performed. We found that the mean age at diagnosis and the percentage of males has not changed much over the years. On the other hand, the duration to diagnosis has shortened; this is particularly evident in the study by Hulka et al. [6]. Another interesting observation is the decrease in the percentage of pyloric tumors palpated, which seems to parallel the increase in the earlier use of UGI series and ultrasound in particular. Again, this is most evident in the study of Hulka et al. [6]. A decline in the postoperative stay was also noted. Unfortunately, no single definition was used to define alkalosis and hypochloremia. Although we noted a trend of improvement over the years, data are not available from older studies. The only exception is the relatively recent investigation by Smith and collaborators [12], where over one-third of the patients had alkalosis, but that study does not include data on imaging.

Discussion

There is an ongoing debate concerning the most efficient and cost-effective algorithm that will lead us to the diagnosis of HPS. The debate is especially relevant in view of the apparent change in the clinical picture [6–8,11–14]. This may be due to the increased accessibility of ultrasound diagnosis [6,7,13] and UGI studies and/or the atrophying clinical skills of pyloric tumor palpation, particularly in young pediatric residents [7,10]. In either circumstance, a variety of methods has been proposed to improve the diagnostic algorithm for HPS. Studies have focused on common biochemical values [8,11,12], gastric aspirate [13], and anatomic anomalies [15] as aids in diagnosis.

We found that certain manifestations in our cohort were similar to the classical earlier descriptions of HPS. These included a predominance of male patients, age of presentation between 3 and 6 weeks with an average age of 40 days at diagnosis, and a significant number of firstborns. Interestingly, we observed a relatively high percentage of firstborns in girls, which was not previously reported.

The common symptom of projectile vomiting was absent in one-third of our cases and peristaltic waves were rarely noted on physical examination. In addition, the pathognomonic pyloric tumor was palpated in only one-half of our patients. This might be a result of declining physician's skills, also noted by others [6,10,16,17], or a result of an earlier presentation.

Recent studies comparing length of illness have noted a shorter duration of symptoms before diagnosis when compared to earlier time periods [6]. Our mean length of illness was 8 days and was similar to recent data reported by Hulka and team [6] and Macdessi and Oates [10]. This contradicts the fact that the mean age at diagnosis is quite consistent over the years [6,8,10,16,18,19]. The probable explanation for this apparent discrepancy lies in the fact

UGI = upper gastrointestinal

that the majority of newborns regurgitate during the first weeks of life, and therefore it is difficult to accurately date the onset of pyloric stenosis symptoms.

Prematurity was noted in 10% of patients, a relatively high percentage. The incidence reported in other large series [6,20] was 3.1–4%. However, all of our premature babies were 35 weeks or older.

Electrolytes and blood gases have been suggested as an aid in diagnosis [1,11]. Oakley and Barnett [11] argue that biochemical values have a predictive value in the clinician's diagnosis of HPS. They presented a model using pH, chloride and base excess with a positive predictive value of 88%. Furthermore, Smith et al. [12] demonstrated that serum bicarbonate levels of $\geqslant$ 29 mmol/L and serum chloride levels of $\leqslant$ 98 mmol/L had a high positive predictive value in identifying patients with HPS. They also note specific cases that were diagnosed with HPS only when a high bicarbonate level was discovered. Overall, metabolic abnormalities in our study were not commonly seen, a finding noted in several other studies [6,7]. Papadakis and co-workers [8] found laboratory abnormalities in only 12% of their patients. We recognized that chloride and bicarbonate levels might be helpful for diagnosing HPS, particularly in the subset of patients presenting without projectile vomiting [Table 1]. These patients demonstrated a significant difference compared to patients who presented with projectile vomiting. One might assume that these differences relate to a longer delay in diagnosis but we found no difference between these groups. No differences in chloride and bicarbonate values were noted for patients presenting without a pyloric mass.

Ultrasound has become the imaging procedure of choice. It is non-invasive, safe, reliable, and accurate in the hands of an experienced sonographer. It is also less costly and time consuming as compared to upper gastrointestinal series [7,10,13]. Van der Schouw et al. [16] demonstrated the advantage of sonography for improving diagnosis of pyloric stenosis. They reviewed 105 sonograms of infants with suspected HPS who had no palpable mass and found that 57% of them had HPS. The correlation between ultrasonographic measurements of length and thickness of the pylorus and the operative findings are encouraging. The fact that pylorus length correlated with length of symptoms may suggest an ongoing process that is developing over time.

Undoubtedly, a thorough history and physical examination should be the first assessment of a vomiting child. Although the literature does not support the argument that the use of imaging and laboratory studies leads to an earlier diagnosis, it may lead to a shorter postoperative stay. In the subgroup of infantile HPS patients with a non-classical presentation (mainly non-projectile vomiting), using imaging and laboratory studies may lead to earlier diagnosis, surgery and discharge.

We conclude that the clinical presentation of HPS has not actually changed despite the easier accessibility of GI imaging studies. The one significant change is the low percentage of pyloric tumors palpated. This may be one of the pitfalls of modern medicine, where the tendency is to rely on technological advances at the expense of clinical expertise.

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Correspondence: Dr. R. Shaoul, Head, Pediatric Day Care Unit, Dept. of Pediatrics, Bnai Zion Medical Center, P.O. Box 4940, Haifa 31048, Israel.
Phone: (972-4) 835-9662
Fax : (972-4) 837-1393
email: shaoul_r@012.net.il