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Perspective

PYLORIC STENOSIS OF INFANCY-PERSPECTIVES Different views from different bridges

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Abstract

Introduction: The differing experiences of the surgeon, he parent and the baby are discussed in terms of what is presently known about the pathogenesis of pyloric stenosis of infancy (PS).

Methods: The experiences are culled from personal experience and from the comments made on online pressure and support groups for pyloric stenosis of infancy. These comments are reviewed from the standpoint of the Primary Hyperacidity pathogenesis of cause.

Conclusion: The present perspectives of the PS surgeon and the difficulties experienced by the PS family fit well which the basic tenets of the Primary Hyperacidity theory. Reference to this theory allows a satisfactory explanation for difficulties in diagnosis and for the variability of presentation.

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INTRODUCTION

The Surgeon

Everybody knows: For progressive pyloric stenosis of infancy (PS), pyloromyotomy reigns supreme! Impending catastrophe transformed safely, in a few minutes, into an enduring cure. A divided incompetent sphincter, now immediately wide open with no further contraction possible. A potentially fatal mechanical problem with a life-saving, mortality-free mechanical solution. Simple and safe.

A broader perspective: In 1921Dr. John Thomson, the acknowledged father of paediatrics in Scotland, first raised the possibility that the presentation and treatment might not be so straightforward. He proposed that the pathogenesis was a continuum-with a broad rather than a narrow presentation. After an analysis of 100 PS babies in the preceding 25 years, he proposed pylorospasm and work hypertrophy as the cause although no primary cause for hypertrophy was defined [1].

He identified *mild* cases of PS.

According to Dr Thomson the mild cases were *not at all uncommon*-they slipped in and out of PS. With modest treatment such as reduced frequency of feeding [2], or indeed no treatment, they would sometimes self-cure and remain cured. The less common acute and ordinary PS was different matters with more severe progressive symptoms which demanded urgent surgical treatment.

In 1961 this theme was further developed by Dr Jacob. He again recognised the milder forms-those in which a long- term cure could similarly be achieved without surgery. A 1% mortality in 100 patients was achieved with the usual medical measures of which a reduced feeding regime was judged to be especially important. 100 more acute cases were also treated surgically by Dr Jacoby personally! and the same mortality achieved(2).

In 1952 McKeown from Birmingham U.K. after an analysis of 1059 PS babies over the previous 10 years, was moved to comment that the presentation was rarely sharp [3]. It could come and go. Many experienced paediatricians will have cause to agree with this comment.The continuation of the Birmingham study also revealed that a post-birth environmental factor was necessary-and that factor was over frequent feeding [4].

The Parent

Multiple on-line support groups have sprung up in recent time. These include *pyloric stenosis support group* on Facebook with 2475 members. The comments regularly attest to unacceptable delays in diagnosis and treatment.

Two common misdiagnoses are offered to the parents of the milk vomiting neonate by the primary practitioner.

1. An allergy; incompatibility or hypersensitivity to the breast or formulae feeds. The evidence to support the existenceof this condition in the first few weeks of life appears to have little if any basis in fact. It will not be discussed further.

2. Mild infantile reflux otherwise known as posseting or spitting. This condition is very common and, without a hiatal hernia, is not associated with systemic symptoms. It invariably self-cures with time. Invasive confirmatory investigations are rarely performed. (The more serious and much rarer form where there is a hiatal hernia and systemic and local symptoms requires invasive investigations and possible surgery. It will not be discussed further).

Both of these mistaken diagnoses will self-cure within a matter of weeks without surgery. They are rarely progressive.

Thomson's milder cases of PS also may self-cure with time. The primary medical practitioner in these instances, will be unaware that he has failed to diagnose a baby with PS.

It goes without saying that PS babies will suffer from reflux. It is obvious. But do babies with reflux suffer from PS?

The late Dr. Isabella Forshall (later President of British Association of Pediatric Surgeons (BAPS)) addressed this issue. Of 58 cases of babies with "lax oesophagus" -(babies diagnosed with reflux with a normally placed cardia)-19% had visible gastric peristalsis and 14% required pyloromyotomy [5]. With hiatal hernia refluxers, only 5% had visible peristalsis and 5% required pyloromyotomy. In this series the "lax oesophagus" babies had an incidence of PS 20 times greater than expected.

It has been argued elsewhere that this is because the pyloric sphincter contraction caused by temporary peak hyperacidity in normal development is common to both simple reflux and PS [6]. The vomiting/regurgitation in both will reflect a functional gastric outlet hold-up when feeds are continued.By its relaxing effect on the lower oesophageal sphincter(LES) acidity facilitates vomiting [7]. Thus, common sense dictates that ultra-sonic sphincter assessment be part of the diagnostic work-up of the apparently refluxing baby.

The Baby

The *PS support groups* and *blogs (https://survivinginfantsurgery.wordpress.com)*from Dr. Fred. Vanderbom testify to the continuing problems post-surgery. One particularly arresting account comes from Lou Cook in an account entitled Unearthing (https://wp.me/plbp5H-MG). Whether imagined or real the experience appears to have been truly unsettling.

Problems with the abdominal incisions -either right sub-costal or vertical continue to be experienced.The scar becomes sensitive to touch and to the psyche. It is to be hoped that the modern minimally invasive approaches will provide a remedy.

The longer-term problems due to continuing hyperacidity in later life have been well documented [8].

Indeed from the accounts in online support groups and blogs, post-pyloromyotomy patients continue to suffer from unexplained abdominal pain and indigestion for which no satisfactory explanation is regularly offered other than "adhesions". Without associated signs of intestinal problems such a diagnosis can only be speculation.

It would surely not be amiss to consider a diagnostic trial of acid-blocking drugson these occasions.

PERSPECTIVES IN PATHOGENESIS

The continuum approach and "rarely sharp" presentation must reflect a changing balance between *promoters* of sphincter contraction/hypertrophy, and *reducing factors* that act to reduce gastric outlet obstruction.

PROMOTERS

Hyperacidity: It is the most potent stimulus to sphincter contraction and hypertrophy [9,10]. The evidence for its primary importance is simply overwhelming.

Babies with PS are hypersecretors of acid both before [11], and after successful pyloromyotomy [12].

Creating hyperacidity produces PS in puppy dogs [13].

Any vomiting baby within the PS age range who is alkalotic (and thus losing more acid than other babies) will have PS [14].

Alkalosis pre-operatively is most quickly cured by acid blocking drugs-again indicating continuing hypersecretion of acid. By these means surgery is not unnecessarily delayed and safer anaesthesia is possible [15].

Non-PS preterm babies who, for good reasons, have indwelling nasogastric tubes have had their acid secretion tested. The male babies secrete more acid than matched females [16].

The 5/1 male pre-ponderance inPS babies is precisely reflected in the male pre-dominance in adult duodenal ulcer patients-a condition known to depend on hyperacidity. They also share a predominance of Blood Group O [17,18].

PS babies treated by cimetidine (an H2 receptor blockers are cured in 17/18 cases when the sphincter diameter is 4 mm. or less) [19].

Retained acid behind a closed pylorus, has nothing to do with it!

The specific mechanism by which hyperacidity provokes sphincter contraction remains unclear. Cholecystokinin release (CCK) released by acidity provokes contraction and prostacyclin again released locally by acidity appears also to cause contraction. Since prostacyclin reduces acid secretion a local negative feed-back system may be operating [20].

Inappropriate over feeding is another significant promoter of sphincter hypertrophy.

The mixing phase when the feed is homogenised prior to exiting the stomach, creates the most frequent contractions with the greatest amplitude [21]. The PS baby is classically described as a vigorous feeder and it would not be surprising if he is over fed. Reduced feeding is an important part of conservative treatment.

Gastric-outlet obstruction (GOO) itself causes an increase in acid secretion by factors which may depend on the elevated gastrins due to antral distension with buffered milk [22].

Neonatal physiology and PS

At birth the infant stomach contains amniotic fluid and is usually alkaline. A temporary wave of acid secretion follows immediately after birth in humans and lasts 24 hours [23]. A

maternal factor transferred from mother to baby which caused foetal acid secretion was proposed. During labour, gastrin is transferred from the mother's placentato the foetus in dogs and causes acid secretion [24,25]. Simplystated, this placental gastrin boost ensures there is acid in the newly born stomach when it cannot yet produce acid itself.

After this in all normal babies who feed, there is a progressive and marked increase in fasting neonatal gastrin from Day 1 to Day 4 to reach levels higher than those in adults [28]. In the same period acidity is increasing until about Day 17 [28], when acidity and gastrin peak for a short time. Thereafter both gastrin and acid fall (Figure 1).

Sequential analysis of neonatal fasting and post-feed gastrins has shown that in the first weeks the fasting gastrins are high and there is no expected post-feed increase.At the age of 2 months the fasting gastrin falls and a post-feed increase occurs. The authors propose that an early insensitivity of the negative feed-back between gastrin and protein stimulation at the antral mucosa level is the explanation [27].

One other explanation is that the negative feed-back between antral acidity is not working in the first few weeks of age. From the moment of birth, acidity will change from neutrality or alkalinity to an acid state at Day 4. From Day 1 to Day 4 there is a huge increase in the fasting gastrin. Thus, gastrin and acidity are rising together as in the Zollinger-Ellison syndrome-and the negative feed-back is clearly not working [28]. When early feeds do not produce a gastrin response and later feeds do, it may simply reflect the buffering effect of feeds on acidity. Early post-feed reduced acidity having no effect on an already maximally stimulated gastrin-and later feeds producing a response when gastrins are lower and under negative feed-back control.

The observation that a post-feed gastrin response is first detected at 2 months of (with a fall in fasting gastrin) is entirely consistent with an initially insensitive gastrin/acid negative feed-back also maturing at that time.

Should the insensitivity also be true for the gastrin/acid negative feed-back other things would follow?

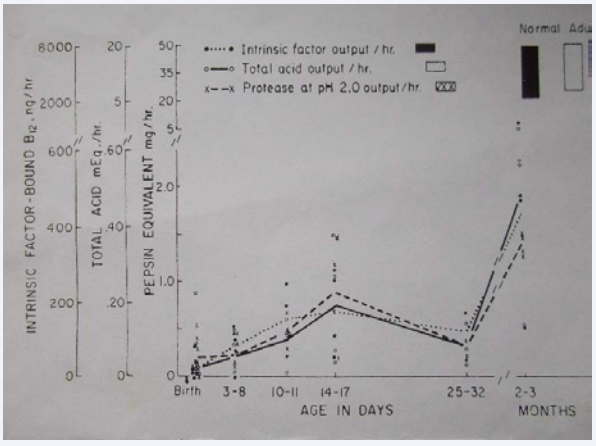

Figure 1 Acid, intrinsic factor and pepsin reach a temporary peak at 17 days.

Firstly, since both gastrin secretion and acidity will be unrestrained in the early days. Gastrin secretion and acid secretion would be unrestrained by one another. They would rise progressively -and they do! [26,28].

Secondly, since both are unrestrained and being maximally secreted (according to the capacity of the neonate), no other stimuli will increase them further.The pentagastrin stimulated acidity on Day 1 would be expected to be the same as the fasting acidity – and it is! [29].

Thirdly, a temporary peak acidity will be anticipated when the negative feed-back is being established Figure 1 shows this to be also true.

The functional consequence of this early maternal boost to acidity and the rising neonatal acid secretion is that the baby isprotected from enteric infections in the early weeks.

One unanticipated consequence is hat the baby who inherits hyperacidity is especially vulnerable to frequent acid-stimulated sphincter contractions and hypertrophy from 2-3 weeks onwards and not before. The delayed presentation at 4 weeks is thus only to be expected. Thus *developmental hyperacidity*, *inappropriate over frequent feeding* and the degree of *gastric outlet obstruction* by the developing PS are the main promoters.

REDUCING FACTORS

Developmental fall in Acidity with time

From around 3 weeks of age both gastrin and acid secretion would come under mutual control and acidity would fall.

Widening of the Pyloric Lumen with age

As age widens the lumen milk feeds are better able to pass through and the acid-provoking element of GOO is lost. Once survival is assured beyond a certain age, there will be no reoccurrence.

Less automatic feeding

Feeding arguably may become less regular and more under the control of appetite. A more experienced and less anxious mother may be better able to avoid inappropriate overfeeding.

The fate of the baby with mild and recoverable PS will depend on the relative strengths of the promoters and reducers. A period of reduced feeding or overfeeding will be sufficient to make the signs and symptoms come and go. The presentation in these mild cases will rarely be "sharp".

CONCLUSION

The differing perspectives of surgeon, parent and baby are analysed in terms of what is known about the Primary Hyperacidity theory of pathogenesis. The observed difficulties in diagnosis such as the common misdiagnosis of reflux; the late presentation of symptoms; the possibilities of self-cure the "unsharp" presentations find an explanation within the theory.

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