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Non-communicating hydrocephalus due to congenital aqueductal stenosis in an infant presenting as an irritable baby syndrome

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A case is reported of an infant with a definitive diagnosis of hydrocephalus secondary to congenital stenosis of the Sylvan Aqueduct who presented to a chiropractor with colicky, irritable type symptoms. These symptoms ultimately proved to be unrelated to the raised intracranial pressure and were ameliorated by the application of one chiropractic adjustment. The literature is reviewed and a discussion of the usual clinical presentation, natural history and pathogenesis of hydrocephalus in infancy is given. The inferential value of this case is to highlight the need for all clinicians engaged in the clinical care of children to perform serial measurements of the head circumference, height, weight and anterior fontanelle as a matter of routine. This case also highlights the fact that alterations of the symptomatic state by a trial of therapy is an unreliable indicator of positive clinical progress.
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KEY WORDS: chiropractic, paediatrics, manipulation, hydrocephalus, colic.

On rapporte le cas, présenté à un chiropraticien, d'un nouveau-né pour lequel a été établi un diagnostic final d'hydrocéphalie due à une sténose congénitale de l'aqueduc de Sylvius, avec des symptômes à type de colique de type irritant. Il s'est finalement avéré que les symptômes n'étaient pas liés à la pression intracrânienne élevée, et l'intervention de chiropraxie les a améliorés. La documentation est révisée, accompagnée d'une discussion de la présentation clinique habituelle, de l'histoire naturelle et de la parthénogénèse de l'hydrocéphalie en bas âge. Ce cas permet, par déduction, de souligner la nécessité pour tous les cliniciens engagés dans les soins cliniques aux enfants, d'effectuer systématiquement des séries de mesures du tour de tête, de la taille, du poids et de la fontanelle antérieure. Ce cas permet également de souligner le fait que des modifications de l'état symptomatique par un essai de thérapie n'indiquent pas avec certitude un progrès clinique positif.
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MOTS CLÉS : chiropraxie, pédiatrie, manipulation, hydrocéphalie, colique.

Introduction

This paper presents a case of non-communicating hydrocephalus, present at birth but not clinically detected until the 12th week of life. The infant was brought for chiropractic care due to persistent and progressively worsening irritability.

In terms of clinical presentation, the signs and symptoms of infantile hydrocephalus are those which accompany raised intracranial pressure, the key features being;

- Accelerating head circumference
- Full or bulging anterior fontanelle
- Widely separated sutures
- Sunset phenomenon ('setting sun sign')
- Irritability
- Failure to achieve developmental milestones[1,2]

The sunset phenomenon occurs when raised intracranial pressure causes declination of the long axis of the eye giving the appearance of the iris as the setting sun and the lower eyelid as the horizon.

In older children, even more overt signs such as seizures, headache, gait disturbance, and decreased visual acuity may be evident.[3]

Remarkably, some patients with hydrocephalus due to aqueductal stenosis are symptom free until late childhood and some will remain asymptomatic throughout adult life.[3,4]

Laurence and Coates report the results of a study of 239 cases of hydrocephalus seen at the Hospital for Sick Children, London over a 20 year period (January 1938 – December 1957). Of the total number of cases followed, 182 were managed conservatively without operation.

The outcome of these cases showed a fatality rate in childhood of 49%. There was also a natural arrest rate of 46% in the preschool age. Of the survivors, however, many showed the classical features of brain damage.[3]

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Gamstorp (1985) reports mortality from all causes of hydrocephalus which is not treated by surgical intervention to be as high as 75%[1] while Lorber and Zachary (1968) describe non-surgical, conservative management as "useless".[6] In addition, Lindsay et al. (1991) describe the morbidity as "significant" in non surgically managed cases.[7]

It has been suggested, however, that such non-surgical management may be appropriate in "simple hydrocephalus" which is uncomplicated by either anatomical malformation or pathology.[8]

While a small number of cases of hydrocephalus in infancy may be described as "simple", by far the dominant cause is impaired circulation or absorption of cerebrospinal fluid.[1]

An obstructive lesion due to anatomic malformation (i.e. Aqueductal Stenosis, Dandy-Walker Syndrome, Arnold-Chiari Malformation), sequelae to an inflammatory process (i.e. meningitis), cerebral tumour, impaired resorption due to membrane damage or increased venous pressure[1,9] are the most common causes of the impaired circulation or absorption.

Case Study

Initial Presentation

The presenting patient was a twelve week old female caucasian, brought to the clinic by the mother. At history, while the mother expressed much frustration and reported being exhausted, she was quite lucid and detailed with her answers to questions and was generally regarded as a reliable historian.

The symptomatic picture was one of prolonged paroxysms of crying accompanied by fisting and "pulling the legs up". The periods of crying had become longer and more frequent with time. The mother complained that she had much difficulty getting the infant off to sleep and when sleep did occur was frequently interrupted after 20 to 30 minutes when the child would awake again crying and pulling its legs up.

Breastfeeding had become increasingly difficult and frustrating due to constant "pulling off" the nipple associated with an extensor contraction posture and screaming.

Initial Examination

On examination, this infant had a head circumference measurement of 44 cm (> 98% + 3SD), a mildly positive setting sun sign and a positive "cracked pot sign". The fontanelle diameter was 4 cm (> 2SD).

MacEwen's cracked pot sign is elicited by placing the ear over one side of the head whilst percussing the other side. In the presence of hydrocephalus the resultant sound is the same as one would get from performing the same procedure on a piece of cracked pottery.

The remainder of the neurologic examination was unremarkable and the vital signs were within normal limits.

The chiropractic biomechanical examination revealed spinal motion unit dysfunction at C1/C2.

Plotting previously taken measurements of length, weight and head circumference was most revealing. Previous measurements were taken at birth, 4, 6 and 9 weeks of age in addition to the measurements taken at presentation (12 weeks). The length and weight measurements were all < + 1SD while the head circumference measurements began at the 50th percentile at birth and progressed to + 3SD by the 9th week. The measurement taken at presentation was > + 3 SD (see Figure 1.)

Diagnosis

A tentative diagnosis of hydrocephalus was made in the child, and in addition the mother's head circumference was measured. When plotted on the adult head circumference percentile charts it was < – 1SD. Since the father was not present at the examination, he was requested to come in the following day to have his head circumference measured and plotted.

Management

The proper protocol for management of patients with previously undetected hydrocephalus calls for prompt referral for medical consultation.

Since the presence of hydrocephalus does not represent either an absolute or relative contraindication to chiropractic spinal manipulative therapy,[10] with respect to the relief of irritability in colicky infants, the C1/C2 dysfunction was corrected using a toggle recoil procedure without the mechanical assistance of a drop headpiece.

The patient's mother was then instructed to bring the child plus the father back the next day. The clinical plan was to examine the father's head in an endeavour to establish whether the child's head circumference was largely due to familial reasons.

Clinical Outcome

The father did not return with the infant for a period of six days. When assessed, his head circumference was > + 1 < + 2 SD and the head circumference of the infant had increased a further 1 cm (see Figure 2).

The symptomatic state of the infant, however, had undergone remarkable change for the better. Both parents reported that the child was now breastfeeding normally, sleep was readily forthcoming and uninterrupted for up to 5 hours at a time. The crying/irritability had completely ceased.

The infant was referred for cranial investigation and subsequently shown by ultrasonography to have congenital stenosis of the Aqueduct of Sylvius. Surgery for insertion of a shunt was performed.

Discussion

This case highlights the need for clinicians to recognize the signs of progressive hydrocephalus and make the appropriate referral for investigation as early in the course of the condition as possible. It further emphasizes the importance of basing a diagnostic impression on objective data rather than symptomatic response to a therapeutic trial.

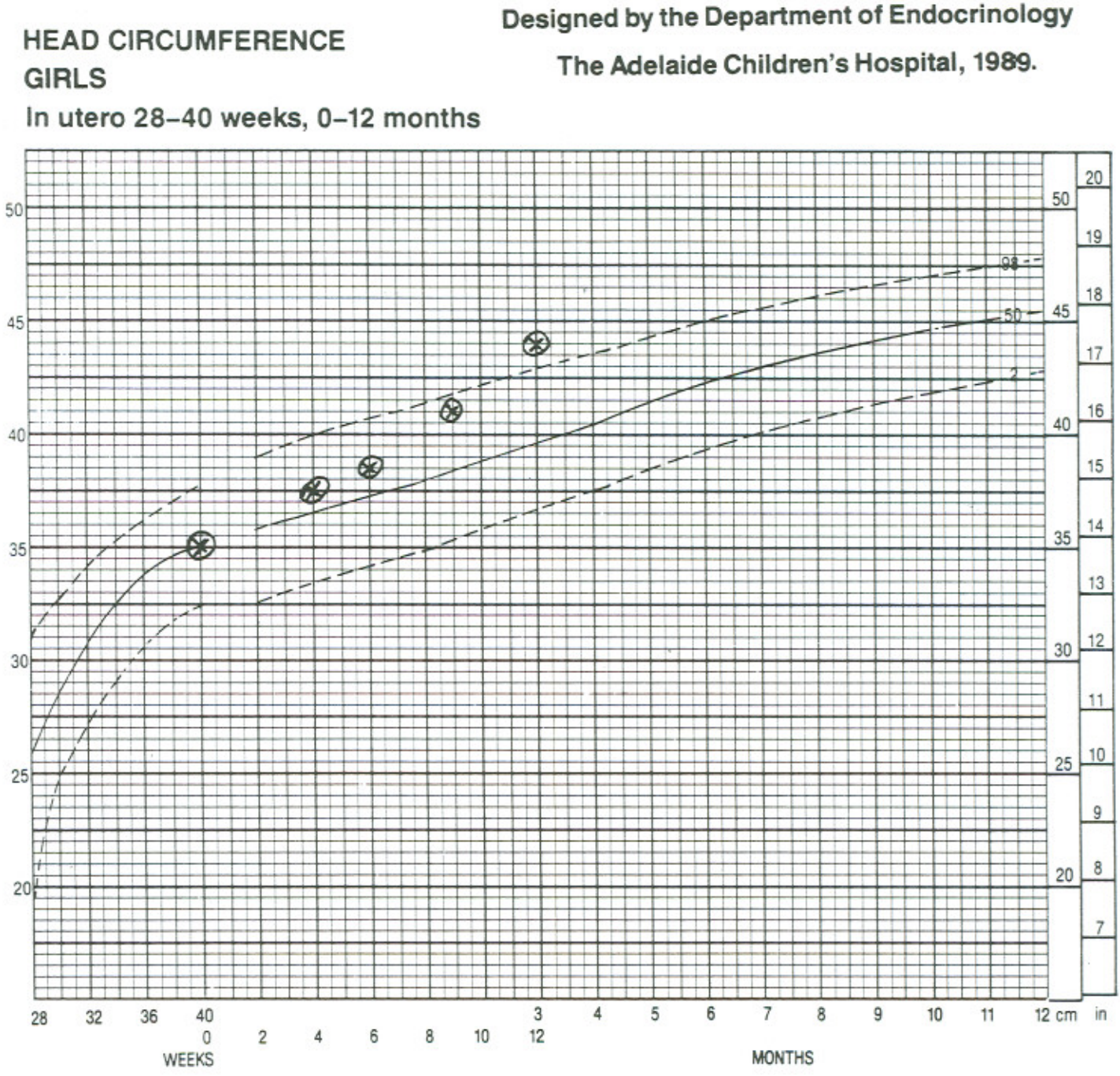

Figure 1 Head circumference percentiles from birth to presentation.

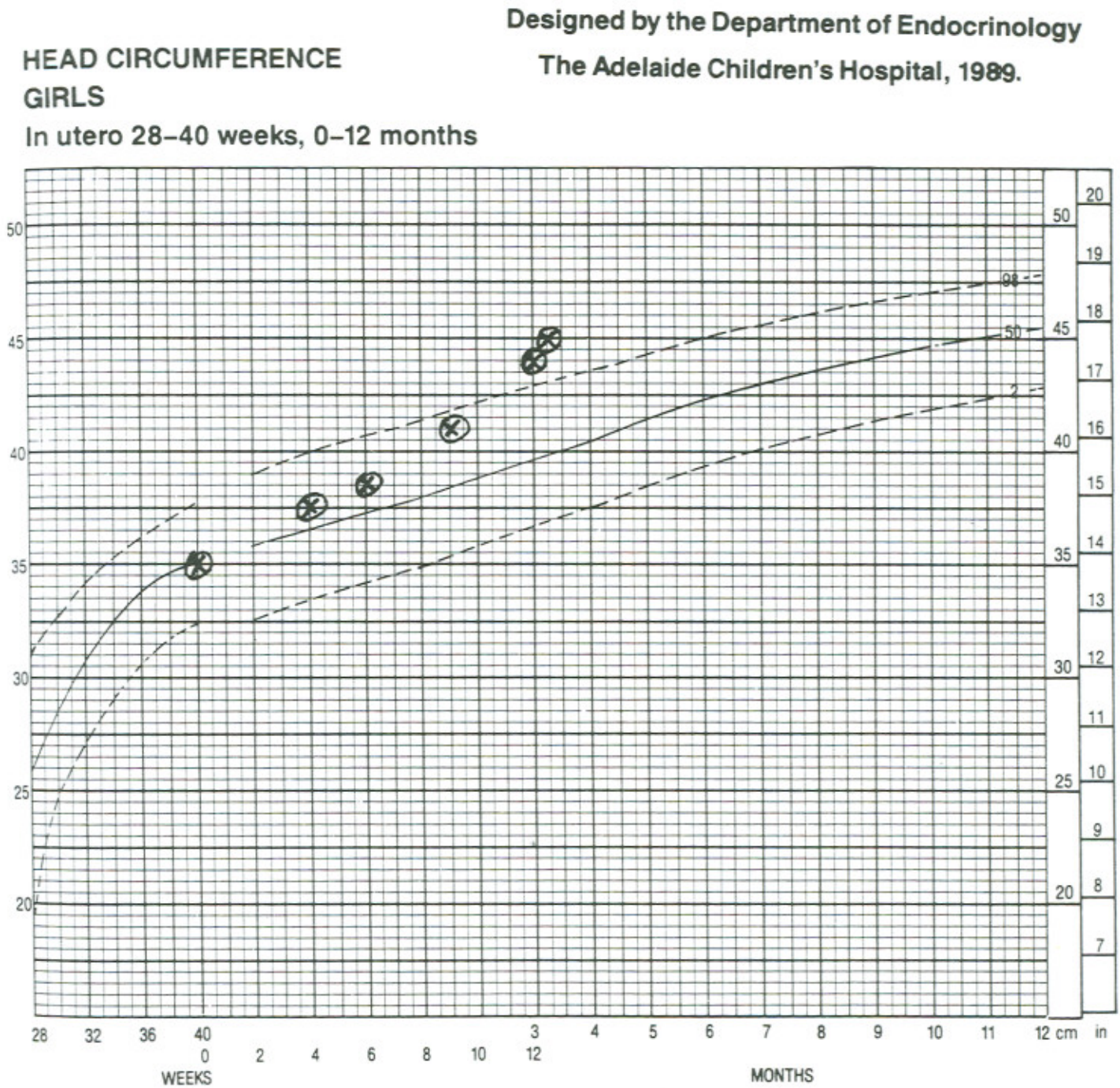

Figure 2 Head circumference percentile on second presentation demonstrating rapidly enlarging head.

The amelioration of the colicky, irritable symptoms by the application of one chiropractic adjustment is consistent with results reported in the literature by Kloughart et al. (1989).[11] While the symptoms in this patient are consistent with those expected in a case of raised intracranial pressure in an infant, in this case they were clearly due to the presence of the spinal motion unit dysfunction, indicating how tenuous it is to rely upon symptomatic change as an indicator of positive clinical progress, when there are co-existing conditions (i.e. hydrocephalus).

A further principle of clinical practice which may be derived from this case is that since serial measurement of the head circumference is a very early and sensitive indicator of the presence of hydrocephalus, such measurements should be taken and plotted on standardised percentile charts by all primary contact clinicians engaging in the clinical care of small children.

From the stand point of the chiropractor in general practice, differential diagnosis is essentially a matter of deciding whether the previously undetected macrocephally is due to hydrocephalus or is in fact normal for that individual. This is accomplished by finding answers to the following questions:

1. Have serially taken measurements of the head circumference been consistently represented at the same standard deviation on the centile charts?
2. Does either parent exhibit a large head?

Positive answers to the above are most comforting, especially when there are no other neurological signs present. Head circumference centile charts for adults are now available.[12]

Summary

Hydrocephalus in infants requires urgent referral for specialist investigation – usually ultrasound followed by computed tomography. Early intervention is now considered essential to either correct the underlying anatomic malformation, insert a shunt to drain the ventricular system, or both.

Diagnosis in general clinical practice is usually made by demonstrating expanding head circumference. This is accomplished by serially measuring and plotting the head circumference on standardised percentile charts. Such anthropometry is both simple and accurate and should form an essential part of the assessment of an infant by all health care professionals in an effort to enhance early detection.

References

1 Gamstorp I. Pediatric Neurology 2nd ed. London: Butterworths 1985.
2 Weiner H, Bresnan M, Levitt L. Pediatric Neurology for the House Officer 2nd ed. Baltimore: Williams and Wilkins 1981.
3 Swaimank. Pediatric Neurology Principles and Practice. Toronto: Mosby Co. 1989.
4 Robinson M. Practical Pediatrics 2nd ed. Melbourne: Churchill Livingston 1990.
5 Laurence K, Coates S. The natural history of hydrocephalus. Arch Dis Child 1972; 61:165.
6 Lorber J, Zachary R. Primary congenital hydrocephalus. Arch Dis Child 1968; 43:516.
7 Lindsay K, Bone I, Callander R. Neurology and Neurosurgery Illustrated 2nd ed. London: Churchill Livingston 1991.
8 Hagberg B, Naglo A. Conservative management of infantile hydrocephalus. Acta Paediat Scand 1972; 61:175–176.
9 Volpe J. Neurology of the Newborn 2nd ed. Toronto: WB Saunders 1987.
10 Idczak R. Aspects of manipulative therapy. Lincoln Institute of Health Sciences. Proceedings International Conference Manipulative Therapy, August 1979, Melbourne.
11 Klougart N, Nilsson N, Jacobsen J. Infantile colic treated by chiropractors: a prospective study of 316 cases. J Manip Physiol Ther 1989; 12(4):281–284.
12 Bushby K, Cole T, Mathews J, Goodship J. Centiles for adult head circumference. Arch Dis Child 1992; 67:1286–1287.