

Some observations on the pathogenesis of syringomyelia

PK NEWMAN, TR TARENTY, JB FOSTER

From the Regional Neurological Centre, Newcastle General Hospital, Newcastle upon Tyne, UK

SUMMARY The pathogenesis of most cases of syringomyelia remains obscure although a modification of the hydrodynamic theory of Gardner allows a logical surgical approach to treatment. Data are presented confirming a high incidence of traumatic birth in patients with syringomyelia who have a Chiari malformation or basal arachnoiditis, but demonstrating no increase in traumatic birth in patients with the Chiari malformation but no syringomyelia. A traumatic birth may be the factor responsible for creating the potential for syringomyelia in those individuals with the embryological defect of the Chiari anomaly.

“My father . . ., had found out, that the lax and pliable state of a child’s head in parturition, the bones of the cranium having no sutures at that time, was such,—that by force of the woman’s efforts, which, in strong labour-pains, was equal, upon an average, to the weight of 470 pounds avoirdupois acting perpendicularly upon it;—it so happened, that in 49 instances out of 50, the said head was compressed and moulded into the shape of an oblong conical piece of dough, such as a pastry-cook generally rolls up in order to make a pye of.—Good God! cried my father, what havoc and destruction must this make in the infinitely fine and tender texture of the cerebellum! . . . But how great was his apprehension, when he further understood, that this force, acting upon the very vertex of the head, not only injured the brain itself or cerebrum,—but that it necessarily squeezed and propelled the cerebrum towards the cerebellum, which was the immediate seat of the understanding!—Angels and ministers of grace defend us! cried my father,—can any soul withstand this shock?” Lawrence Sterne, in *The Life and Opinions of Tristram Shandy, Gentleman*, 1st published 1759-67, Vol II p 151-152, Penguin English Library.

Syringomyelia is the generic term for a group of disorders which have cavitation of the spinal cord in common. The majority of syringomyelia cavities are associated with the Chiari type 1 anomaly and a smaller group with basal arachnoiditis which is

demonstrated at myelography or surgery. The term “communicating” syringomyelia has been applied to these cavities. Smaller numbers of cavities are related to other pathological conditions, in particular spinal cord tumour, posterior fossa tumour and cyst, and spinal cord trauma. A significant number of patients with clinically evident syringomyelia have entirely normal myelograms. The pathogenesis in this group is obscure, although some may have unrecognised cord tumours or vascular disturbances of the spinal cord. The spectrum of pathological states identified in the Newcastle syringomyelia series is illustrated in fig 1.

The theory underlying the formation of “non-communicating” forms of syringomyelia is the subject of little dissent, but considerable controversy surrounds the pathogenesis of “communicating” syringomyelia. Traditional ideas have been discarded largely in favour of the still disputed hydrodynamic theory formulated by Gardner *et al*¹ and Gardner,^{2,3} and subsequently modified by Williams.^{4,5,6} Gardner’s theory postulates primary failure of normal perforation of the rhombic roof in early embryonic life, leading to a partial obstruction of the outflow of cerebrospinal fluid from the fourth ventricle, with several potential consequences including persistence of a dilated central canal. Dissection of the cord results from constant arterial pulsation transmitted to the ventricular fluid from the choroid plexus, and hence funnelled into the communicating central canal with what Gardner describes as a water-hammer effect, eventually breaching the ependymal lining of the hydromyelic cavity and producing clinical syringomyelia.

Address for reprint requests: Dr PK Newman, Newcastle General Hospital, Westgate Road, Newcastle upon Tyne NE4 6BE, UK.

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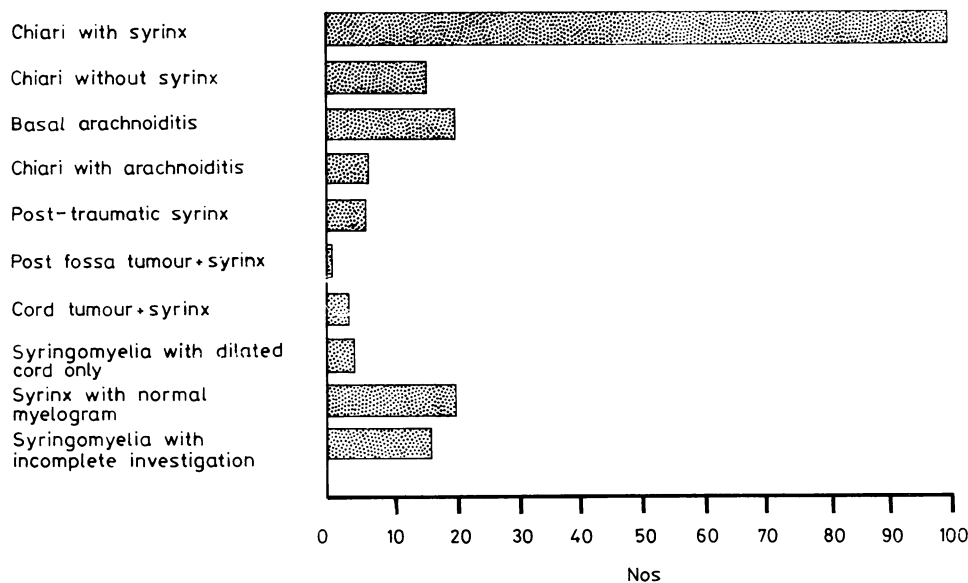


Fig 1 Newcastle syringomyelia series: 1960-1980.

William's modification of this concept rejects arterial pressure pulses as a significant mechanical factor, and instead favours intracranial and spinal, venous and cerebrospinal fluid pressure differentials. Various manoeuvres such as coughing, stooping, straining and voiding, produce increased intrathoracic and intra-abdominal pressure, raising the central venous pressure and distending the spinal epidural plexus. This distension in a confined space is accompanied by a rapid displacement of spinal fluid into the head which is normally followed by the return of the fluid into the spinal subarachnoid space when the pressure falls. In an individual with cerebellar ectopia causing partial obstruction at the foramen magnum, the initial surge of fluid into the head occurs, but as the fluid drains from the head the cerebellar tonsils in the foramen magnum impact in a fashion analogous to a ball-valve mechanism. The patent central canal, at lower pressure now than the cranial compartment, thus becomes the recipient of this trapped fluid and these repeated episodes of cranio-spinal dissociation of pressure gradually enlarge the hydromyelic cavity, with eventual rupture. In this context, it is of interest that a number of our cases of syringomyelia described severe asthma in childhood and early adult life.

These theories have received widespread acceptance and led to substantial changes in the surgical approach to syringomyelia⁷ but many aspects are disquieting, particularly the paucity of pathological confirmation. Central to the hydrodynamic theory is the concept of a patent communication between the fourth ventricle and the central canal and although

only a minority of pathological specimens show this communication,⁸ analysis of 20 cases of syringomyelia at autopsy showed some sort of communication in 17, albeit too small to be of significance in the view of the examiner.⁹ West and Williams¹⁰ have reviewed the evidence from radiological, radioisotope and dye studies, but again communication can be demonstrated by these methods in only a minority of cases.

The concept of central canal patency receives some support from an autopsy study conducted in this department (T Terenty and M Taylor). The spinal cords from 60 patients dying without neurological disease were removed, in continuity with the brain stem where possible, and examined microscopically after formalin fixation. Central canals showed great morphological variability, ranging from an ill-defined core of ependymal cells to a patent canal of up to 1000μ in diameter (fig 2). The results are displayed diagrammatically in fig 3, where each line represents an individual spinal cord with the cords grouped in decades according to the age of the patient. The height above the base line represents the percentage number of cord segments in which the canal is patent, and that below the base line the percentage number in which the canal is obliterated. Thus, in the second decade canals may be patent throughout their length, but in later life the canal is progressively occluded, occasionally remaining patent in the lower cervical or lumbar segments.

In our opinion, it therefore seems reasonable to postulate the existence of a patent communication between fourth ventricle and syringomyelic cleft at

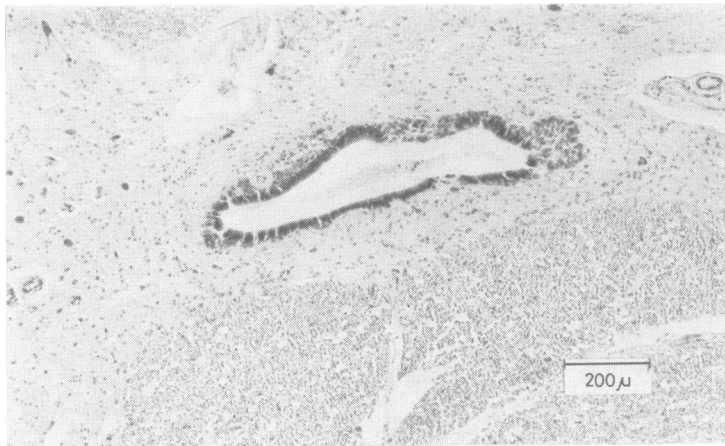


Fig 2 Section through first lumbar segment, female aged 60, showing a patent central canal measuring 800 μ in the transverse plane. This was identified throughout the length of the spinal cord.

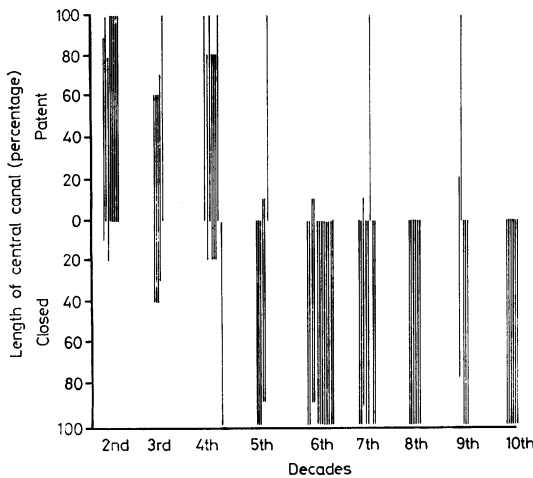


Fig 3 Central canal patency contrasted with age in decades—see text.

an early stage in the development of the syringomyelia, but to accept that in many cases this conduit may partly or completely close in later life. Continuing neurological deficit after this closure can be explained by the persistence of destructive fluid shifts within the dilated cavity.

Foster and Hudgson⁸ observed that birth trauma may have been implicated in cases of basal arachnoiditis and syringomyelia. Other possible aetiological factors, such as meningitis or confirmed subarachnoid haemorrhage, are present in only a very small minority. A subsequent study¹¹ went further and suggested that traumatic birth may be of fundamental importance in the pathogenesis of all cases of communicating syringomyelia. We decided to analyse this apparent association in a series of our

patients with syringomyelia and, moreover, to compare these cases with a group of patients who had the Chiari anomaly but no clinical or myelographic evidence of syringomyelia.

Methods

The birth history of 68 patients with syringomyelia and of 12 patients with an isolated Chiari anomaly was obtained by direct interview or, if this was not possible, by postal questionnaire. Fifty controls, selected at random from routine admissions to a neurological ward, were questioned by the same method. Information was sought about birth weight, birth rank, prematurity and post maturity, abnormal presentation, prolonged labour, instrumental delivery, operative delivery, neonatal disturbance and maternal illness or complication. In approximately one-third of both the study patients and the control group, insufficient information was available, and in these cases birth was recorded as normal. This method of collecting and presenting obstetric information is not ideal, but in the light of the results it can be predicted that more abnormal births will have been missed in the syringomyelia group than in the controls. The syringomyelia and Chiari groups were compared with the controls (fig 4) with regard to the frequency of forceps delivery, prolonged labour and abnormal birth. Abnormal birth was defined as any perinatal disturbance leading to

Birth trauma and syringomyelia: patients	
1	Controls — 50 M:F 22:28 Mean age 45.2 (15-80)
2	Syringomyelia — 80 M:F 38:42 Mean age 46.9 (17-72)
	(i) Chiari with syrinx 43
	(ii) Chiari without syrinx 12
	(iii) Arachnoiditis with syrinx 11
	(iv) Clinical syringomyelia with normal myelogram 14

Fig 4 Birth trauma study: patients.

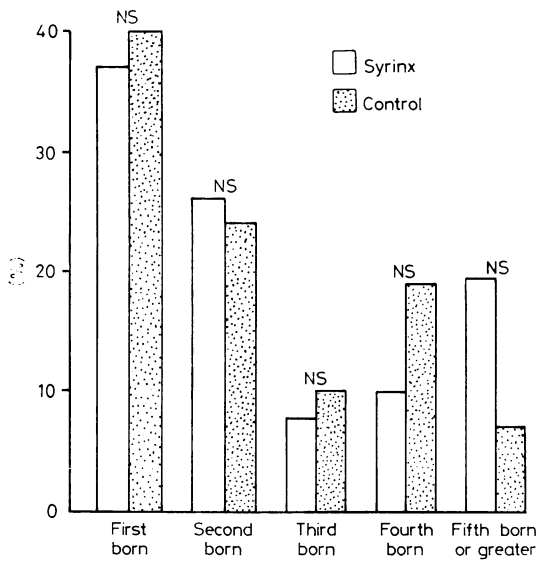


Fig 5 Birth rank.

difficult or traumatic labour, and thus included forceps delivery and prolonged labour, but also other factors such as breech presentation, face presentation, prolapsed umbilical cord and domiciliary maternal anaesthesia.

Results

An insufficient number of the patients knew their birth weight and hence this factor could not be analysed. Birth rank was not different in the syringomyelia cases and the controls (fig 5). No patient in the Chiari group recalled having been told of a delivery by caesarean section, but three patients in the syringomyelia group had been delivered by this method. On two occasions the operation had

Birth trauma and syringomyelia: results			
	Forceps %	Prolonged labour, %	Abnormal birth, %
Control	4/50 (8)	6/50 (12)	9/50 (18)
Total syring	25/80 (31)	24/80 (30)	38/80 (48)
Clinical syringomyelia with normal myelogram	1/14 (7)	0/14 (0)	2/14 (14)
Chiari no syring	1/14 (7)	2/12 (17)	3/12 (25)
Chiari syring	14/43 (33)	16/43 (37)	23/43 (53)
Arachnoiditis syring	9/11 (82)	6/11 (54)	9/11 (82)

Fig 6 Birth trauma study: results.

followed arrested vaginal delivery. The details were not available in the third case. Forceps delivery, prolonged labour and abnormal birth were no more frequent in the Chiari patients without syringomyelia or the "syringomyelia" patients with a negative myelogram, than in the controls. However, the Chiari patients with syringomyelia and the patients with a basal arachnoiditis and syringomyelia were far more likely than the controls to have experienced

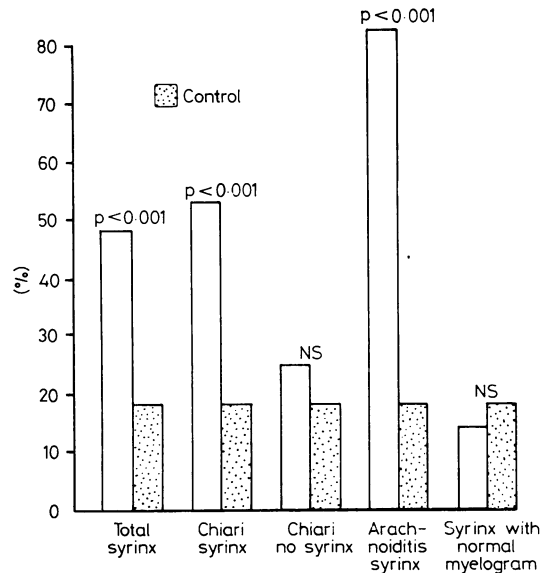


Fig 7 Forceps delivery.

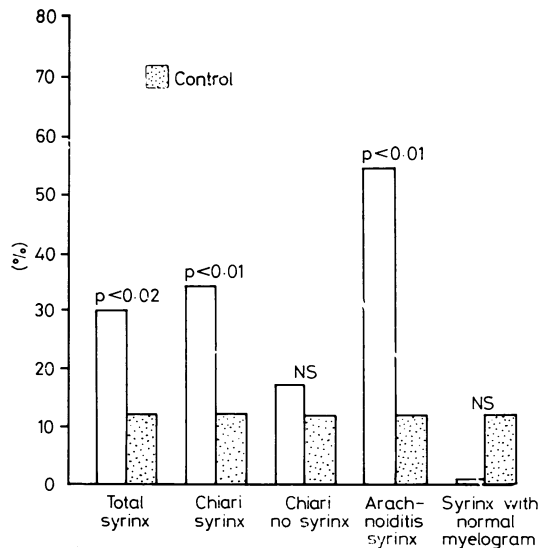


Fig 8 Prolonged labour.

a forceps delivery, prolonged labour and abnormal birth (fig 6). These differences were statistically significant (figs 7, 8, 9). A comparison of the Chiari patients with syringomyelia directly with the Chiari patients without syringomyelia (fig 10) illustrates the differences between these two groups which do not

reach statistical significance because of the relatively few patients in the latter group.

Discussion

Williams¹¹ stated that difficult labour was more common in patients with communicating syringomyelia than in controls, or when these cases are compared with data available from the 1958 Perinatal Mortality Survey. The present study confirms this observation and, furthermore, demonstrates a varying frequency within the different sub-groups of syringomyelia and patients with an uncomplicated Chiari anomaly. It is likely that a difficult birth gives rise to syringomyelia and not the reverse, and there is no evidence that the difficult labour arose because of any putative disturbance *in utero*. It should be stressed that overt hydrocephalus and spina bifida were unusual in this series: there were only two cases with clinically recognisable hydrocephalus and three cases with spina bifida. All three spina bifida patients had the Chiari anomaly but only one had syringomyelia, and it was in this case that a difficult birth was recorded.

We suggest the following sequence of events operating in the formation of a syrinx (fig 11). Partial failure of perforation of the rhombic roof delays termination of the phase of foetal "physiological" hydrocephalus, leads to a persistent and dilated central canal and may be responsible for the hind-

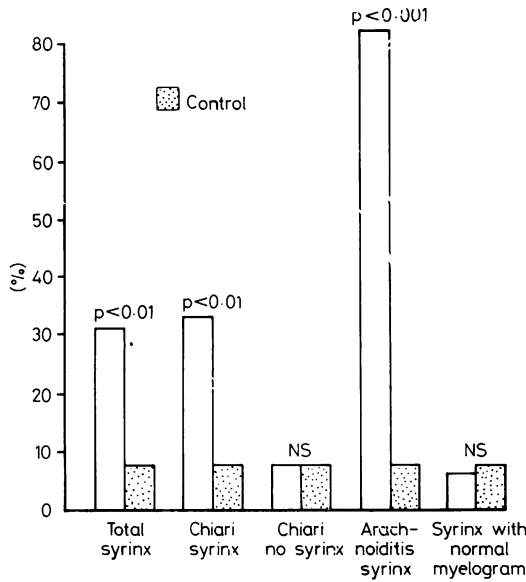


Fig 9 Abnormal birth.

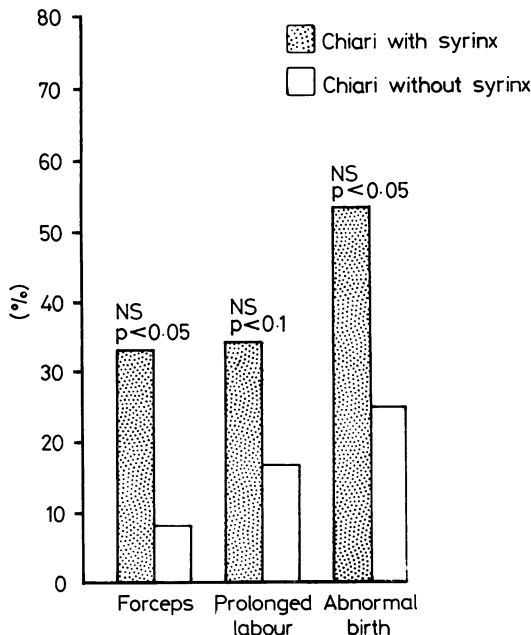


Fig 10 Chiari anomaly with and without syringomyelia.

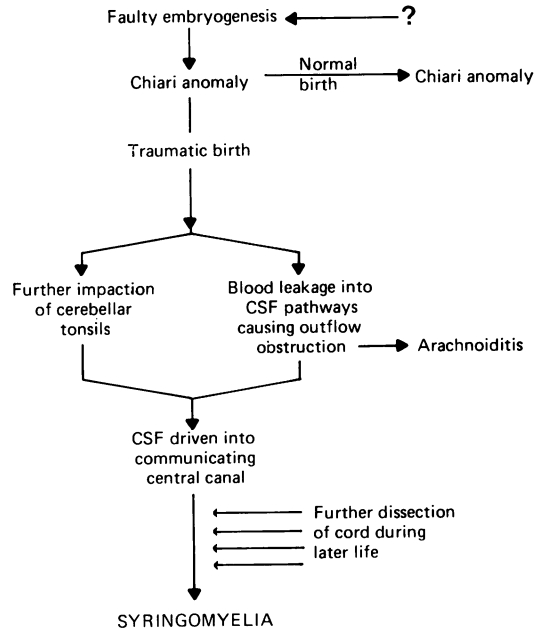


Fig 11 Hypothesis: the pathogenesis of syringomyelia.

brain anomaly. Foster and Hudgson⁸ cite the early work of Weed¹² in suggesting that this fundamental embryological disturbance occurs at the eighth week of foetal life, although a recent study of aborted fetuses with the Chiari malformation was unable to define the sequence of onset of the hind-brain anomaly and its associated hydrocephalus.¹³ If the individual with the Chiari type I anomaly has a normal birth, then the inference from our data is that there may be no progression to syrinx formation. However, if the labour is traumatic, then a combination of prolonged and forceful expulsive pressures in an obstructed birth canal, excessive moulding of the foetal head and aggressive instrumentation may be associated with impaired venous return from the head of the infant. These factors may cause further impaction of the cerebellar tonsils, the ependyma of the central canal may rupture, or there may be traumatic subarachnoid and ventricular haemorrhage further obstructing the flow of spinal fluid and causing the basal arachnoiditis. In such a situation it seems likely that the massive extracranial and intracranial pressures of birth may be reflected in the cerebrospinal fluid pathways, causing irreversible dilatation of the central canal of the cord with the resulting potential for, or actual development of, syringomyelia. In later life the cord cavitation is further expanded, as discussed earlier, and clinically evident syringomyelia results.

A difficult labour is a factor in the pathogenesis of syringomyelia where a Chiari anomaly is present and we suggest that a traumatic birth is probably responsible for the basal arachnoiditis seen in some cases. The above mechanistic account necessarily represents an oversimplification of a complex sequence of patho-physiological events. Superficially, this hypothesis should require a diminishing incidence of syringomyelia in parallel with improved obstetric standards in this country, and a high incidence of syringomyelia in areas of poor obstetric health.

Although there is no evidence of this at present, comparative studies and epidemiological analysis of cases of syringomyelia may be of interest.

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