The main problem with this valve arises when the patient moves upwards and downwards. Such a situation has been simulated by the addition of a pulsating waveform of variable amplitude to the proximal pressure (frequency was controlled from 90 revolutions/min to 5/min). As a result, the lower end of the pressure-flow performance curve was consistently twisted to the left (towards lower pressures, figure (B)). In vivo, variations in intraventricular pressure, produced by either an increased magnitude of the pulse waveform or repeated body movement, may accelerate the drainage rate, in some cases possibly leading to overdrainage. The above phenomenon is probably a common feature of all gravitational valves, which should always be implanted with caution, taking into account the usual risk factors for serious consequences from overdrainage (thin cerebral mantle and a high brain compliance) and the possible lifestyle of the patient after surgery.

Contrary to other gravitational valves, the Chhabra shunt does not have any valve working with the weighting balls that would prevent the reflux of CSF from the peritoneal cavity to the ventricles. Therefore, CSF reflux is possible, undoubtedly in the upside down and also in the horizontal body position (albeit at a lower rate).

The behaviour of the valve in a strong magnetic field (1.5 T) does not exhibit any alarming variation. However, the artefact on MRI may be considerable.

In conclusion, the intentions of the designers of the Chhabra shunt to make it operate at a higher operating pressure in the vertical than the horizontal body position were confirmed during this evaluation. The shunt has a repeatable pressure-flow performance that does not differ from the performance of more expensive valves manufactured by the big western corporations. However, because the shunt has a very low resistance to flow, overdrainage may be a problem depending on the patient's life activity. Reflux of CSF from the peritoneal to the ventricular cavity is possible.

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A case of chronic paroxysmal hemicrania responding to subcutaneous sumatriptan

Chronic paroxysmal hemicrania was first described by Sjaastad and Dale in 1974.¹ It is characterised by attacks of excruciating unilateral oculotemporal pain associated with autonomic changes, such as lacrimation, rhinorrhea, ptosis, miosis, and conjunctival suffusion. It differs from cluster headache for its female predominance, brevity (5–45 minutes), and frequency (5–20/day) of pain attacks, as well as for its response to indomethacin in doses of up to 150 mg/day.² Experience with subcutaneous sumatriptan in chronic paroxysmal hemicrania is scarce. Here we report on the effect of subcutaneous sumatriptan in a patient with chronic paroxysmal hemicrania.

In May 1994, when 34 years old, this previously healthy woman started to have attacks of severe temporal and periorbital pain usually associated with conjunctival injection, lacrimation, ptosis, eyelid oedema, and nasal congestion. Frequency and duration of attacks were variable. She experienced at least five pain attacks a day from the beginning of her clinical presentation, but every few months the frequency increased up to more than 20 episodes each day in bouts lasting 20–30 days. The duration of attacks ranged from several minutes to 1 hour. A diagnosis of cluster headache was made. She was treated with the maximum doses that she could tolerate of verapamil (40 mg/6 h), and methysergide (1 mg/6 h), without success. Ergotamine (2 mg at night) was added with doubtful symptomatic improvement, so domiciliary 100% oxygen (7 l/min) and/or subcutaneous sumatriptan (6 mg) were prescribed. She thought that oxygen inhalation during the attacks did not stop the pain but alleviated it if the episode was mild and if the oxygen was taken during the earliest part of the attack. Subcutaneous sumatriptan stopped the attacks in less than 15 minutes. In addition, during the bouts, when she experienced multiple consecutive pain attacks, the patient noticed that after subcutaneous sumatriptan she was free of pain for a maximum of 8 hours and a minimum of 6 hours. She was sent to us in March 1996. Systemic and neurological examinations, as well as high resolution brain CT (with and without contrast) were unremarkable. Indomethacin (75 mg daily), was started, with absolute effectiveness after 4 days. Indomethacin has been withdrawn twice, with immediate pain reappearance. During more than one year of follow up, she has twice needed to increase indomethacin up to 125 mg daily for several weeks because of further pain episodes, which promptly stopped with subcutaneous sumatriptan on the four occasions that this was injected. She is now taking 75 mg indomethacin daily and ranitidine, with very infrequent attacks of only slight pain.

The differential diagnosis between chronic paroxysmal hemicrania and chronic cluster headache still poses some problems. With that in mind, this woman clearly meets the criteria of the International Headache Society classification for diagnosis of chronic paroxysmal hemicrania.² Subcutaneous sumatriptan consistently relieved her chronic paroxysmal hemicrania attacks in less than 15 minutes and protected her from further pain attacks for 6 to 8 hours. Subcutaneous sumatriptan is a very effective symptomatic treatment for cluster headache pain attacks,

this confirming that trigemino-vascular activation with cranial vasodilatation or plasma protein extravasation are the pathophysiological explanation for cluster headache attacks.³ The aetiology and pathogenesis of chronic paroxysmal hemicrania are still unknown. Goadsby and Edvinsson have recently reported a patient with chronic paroxysmal hemicrania in whom concentrations of calcitonin gene related peptide and vasoactive intestinal polypeptide, two markers of trigemino-vascular activation, were raised in the cranial circulation during attacks. Moreover, the concentrations of both peptides became normal when indomethacin was given, all this suggesting a shared pathophysiology between chronic paroxysmal hemicrania and cluster headache.⁴ The finding of the effect of subcutaneous sumatriptan in chronic paroxysmal hemicrania clarifies this point further. To the best of our knowledge, the effect of subcutaneous sumatriptan has been reported only in three patients with chronic paroxysmal hemicrania and gives discordant results.^{5–7} One patient, a male but with very typical chronic paroxysmal hemicrania attacks, did not respond to subcutaneous sumatriptan on two different occasions, although the duration of this patient's attacks was not mentioned.⁵ Two other patients had a clearly positive response.^{6,7} These two patients, however, were atypical, one also having intracranial hypertension and the other both chronic paroxysmal hemicrania and trigeminal neuralgia. Although more experience is necessary, our findings suggest that, in parallel with migraine and cluster headache, trigemino-vascular activation also occurs in chronic paroxysmal hemicrania.

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IgM anti-GM2 antibody in a patient with Guillain-Barré syndrome subsequent to cytomegalovirus hepatitis cross reacts with N-acetylgalactosaminyl GD1a

Anti-GM2 antibodies are associated with acute cytomegalovirus infection in patients with Guillain-Barré syndrome.^{1,2} Irie *et al*¹ reported that anti-GM2 antibodies do not

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Figure 1 Binding of gangliosides with serum IgMs from a patient with Guillain-Barré syndrome subsequent to acute cytomegalovirus hepatitis. Lane 1: bovine brain gangliosides GM1, GD1a, GD1b, and GT1b. Lane 2: purified GM2 and GalNAc-GD1a. (A) Thin layer chromatography plate stained with orcinol. (B) Chromatogram immunostained after overlaying it with serum from the patient (1:100) then with peroxidase conjugated antihuman IgM (Dako, Denmark; 1:1000). The chromatography plates were developed in a solvent system of chloroform/methanol/ 12mM magnesium chloride in water (5:4:1, volume). The IgMs from the patient reacted specifically with the GM2 and GalNAc-GD1a.

bind to N-acetylgalactosaminyl GD1a (GalNAc-GD1a), a terminal trisaccharide identical to that of GM2. Jacobs *et al*² did not test whether IgM anti-GM2 antibodies react with GalNAc-GD1a. By contrast, Kusunoki *et al*³ reported that a patient with Guillain-Barré syndrome had IgM antibody activity against both GalNAc-GD1a and GM2, but in whom no result of anti-CMV antibody was shown. We had a patient with Guillain-Barré syndrome subsequent to acute cytomegalovirus hepatitis who had IgM antibodies both to GM2 and GalNAc-GD1a. We investigated whether the IgM anti-GM2 antibodies cross react with GalNAc-GD1a.

Three days after an episode of stomatitis, a 32 year old man noticed left sided facial weakness and dysaesthesia distally in his feet. Over a day, the facial weakness extended to the right side and dysaesthesia progressed to the limbs. He developed progressive weakness of the limbs on day 4, and the next day was unable to stand. On day 5, the patient presented with facial diplegia, severe limb weakness, and moderate reduction of superficial sensation distally in all four limbs. The respiratory muscles were slightly affected. Deep tendon reflexes were absent. Liver enzyme concentrations were raised slightly. Protein concentration in CSF was 109 mg/dl and cellularity was normal. High IgM anti-CMV antibody titres were found in both serum and CSF by enzyme linked immunosorbent assay (ELISA). An electrophysiological study suggested that the predominant process was demyelination involving the motor nerves. Right ulnar sensory nerve action potential was absent. Visser *et al*⁴ reported that patients with Guillain-Barré syndrome with associated cytomegalovirus were young and often developed severe sensory loss with facial nerve involvement and respiratory insufficiency. They also reported a strong correlation between cytomegalovirus infection and severe sensory loss. The clinical manifestations in our patient were similar to those reported by Visser *et al*.

Thin layer chromatography with immunostaining showed that the serum IgM from our patient reacted strongly with GM2 and

GalNAc-GD1a, but not with GM1, GD1a, GD1b, or GT1b (fig 1). ELISA confirmed that his serum had high titres of IgM antibodies to GM2 (1:25 600) and to GalNAc-GD1a (1:12 800) on day 4. Plasmapheresis was performed on days 5, 8, 24, 27, and 29, he was able to walk without assistance on day 60, but his facial diplegia showed slow improvement. The IgM anti-GM2 (1:800) and anti-GalNAc-GD1a (1:400) titres in this patient were significantly reduced on day 39. In the absorption study his serum was added to separate wells coated with individual ganglioside antigens (GM2, GalNAc-GD1a, GM1, and GD1a). The absorption rate was calculated from (1-(optical density at 492 nm in the well with serum with absorption treatment)/(optical density in the reference well with serum without absorption treatment)). No IgM anti-GM2 antibody was absorbed by GM1 or GD1a (fig 2). By

Figure 2 Absorption study of serum IgM anti-GM2 antibody from the patient carried out with GM2, GalNAc-GD1a, GM1, and GD1a. Serum samples were diluted serially from 1:250 to 1:8000 and added to wells coated with 5 pmol portions of each ganglioside antigen. The absorption rate was calculated from (1-(optical density at 492 nm in the well with serum with absorption treatment)/(optical density in the reference well with serum without absorption treatment)). The IgM anti-GM2 antibody in our patient was not absorbed by GM1 and GD1a but was by GM2 and GalNAc-GD1a.

contrast, IgM anti-GM2 antibody was absorbed by GalNAc-GD1a. Larger studies are needed to confirm whether (1) some patients with Guillain-Barré syndrome after CMV infection have IgM anti-GalNAc-GD1a antibody and (2) that the anti-GalNAc-GD1a antibodies cross react with GM2.

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Electrophysiological recordings in bilateral hemifacial spasm

Hemifacial spasm typically consists of unilateral involuntary spasms of the mimic muscles innervated by either facial nerve. There is some controversy whether mechanical irritation of the facial nerve entry zone by vessels in the cerebellopontine angle or hyperexcitability of the facial nerve nucleus is the main cause of this problem.^{1,2} To our knowledge, only two cases of bilateral hemifacial spasm have been described in some clinical detail but without electrophysiological recordings.^{3,4} The mechanism underlying the bilateral occurrence of spasms has not been investigated so far: either hyperexcitability of a facial nerve nucleus spreading to the contralateral side or independent mechanical irritation of both nerves might be involved. We report a patient with predominantly left sided spasms, electrophysiological diagnosis of bilateral facial spasm, the clinical course after surgical treatment according to MR evidence of elongated vertebral and basilar arteries, and implications of electrophysiological recordings on the pathogenesis of bilateral hemifacial spasm.

This 60 year old male patient presented with a history of gradually increasing left eye twitching over the past few years. He did not notice involvement of other facial muscles or of associated symptoms such as hearing deficits, vertigo, or headache. Visual inspection showed frequent clonic involuntary synchronous contractions of his left orbicularis oculi and left orbicularis oris muscles. On voluntary contraction of the orbicularis oculi muscle, there was some synkinesis of the left orbicularis oris