Foramen Magnum Decompression in Arnold-Chiari Syndrome Type I and Idiopathic Syringomyelia

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Introduction


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There is an open debate on the diagnosis and types of the Arnold-Chiari Syndrome (ACHS), joined by the discussion about its treatment and that of the frequently associated idiopathic Syringomyelia (ISM). The standard treatment for the first type of ACHS, the Arnold-Chiari Syndrome Type I (ACHSI) and sometimes for idiopathic Syringomyelia (ISM), is the foramen magnum decompression (FMD).

Lately, some neurosurgeons, upon observing the disparity between the risks of the surgical technique of the FMD and those of the conditions that are to be treated, ACHSI and ISM, recommend its application in extreme cases without specifying too clearly what they mean by an extreme case, possibly referring to cases where the clinical expressions are very manifest as a vast majority of cases are apparently asymptomatic.

The existence of compilation publications about the application of the FMD in ACHSI and ISM has recently allowed a statistical assessment of the results and to determine the convenience of the treatment.

After going shortly over the historical aspects, we propose a bibliographical review of the FMD applied to ACHSI and ISM to carry out a comparison of its results to its natural course. The purpose is to validate its usefulness if proven that the mortality rate ascribed to the surgical technique is lower than that of the natural course, considering that the balance between results and complications should be beneficial for the

patient.

Spontaneous mortality in ACHSI and ISM

There is evidence of sudden death in relation to ACHSI/ISM but possibly due to its scarce frequency together with a difficult diagnosis, there are few statistics that contribute to the incidence or prevalence or a sudden death rate in relation to ACHSI/ISM. The majority of publications that make reference to it tend to be of just one case, some of two cases or very few of several cases [2, 36-47].

Every year, 75 million people are born throughout the world according to the United Nations Populations Fund (UNFPA). In 30 years 2.25 billion individuals are born. Taking into account that the incidence of the ACHSI/ISM complex is of 1:1000 births (numbers from the 19th and 20th century), the number of cases with the ACHSI/ISM complex in three decades is of 2.25 million.

According to the retrospective study by Massimi L et al (2012) [2] the total number of cases of sudden death attributed to the ACHSI/ISM complex published in three decades reaches 8 cases out of the 41 cases that presented an abrupt clinical onset, related to a decompensation of the ACHS/ISM complex, rushing towards motor deficit (36.5%), respiratory insufficiency (29%), cranial nerve paralysis (17%) and cardiac arrest (14.5%) among the most frequent manifestations. This confirms that an abrupt clinical onset can have severe and potentially fatal consequences.

Taking this information into consideration, the spontaneous or sudden death rate of the ACHSI/SM complex is of 8/2.25 million patients in three decades = 0.0004%. This theoretical rate is far from real given that an important number of sudden death cases related to the ACHSI/ISM complex go undiagnosed, due to a lack of means, due to ignorance or due to a lack of appropriate health check ups. Despite the circumstances, the real rate would statistically be very low and therefore not much would change when comparing it to the iatrogenic rate of FMD applied to the ACHSI/ISM complex.

Massimi et al attribute sudden death in the ACHSI/SM complex to the acute compression by ectopic cerebellar tonsils and syringomyelia/syringobulbia of the brainstem and of the upper cervical spinal cord, frequently prompted by a minor injury followed by an alteration of the medullary baroreceptors and the reticular substance of the mesencephalon (cardiac arrest, syncope) and of the medullary chemoreceptors with the phrenic nerve nuclei (respiratory insufficiency), and in a lesser way due to the affectation of the nervous nuclei of the cranial nerves (causing cardiac arrest, paralysis of the cranial nerves) and of the pyramidal tracts (with a motor deficit). Around 87% of patients in this review that ended up in the clinical picture of sudden death was asymptomatic prior to the acute clinical onset. This is a relevant fact for treating the ACHSI/SM complex once it is diagnosed.

Historical antecedents of the FMD applied to ACHSI and ISM

The first person to apply the Foramen Magnum Decompression (FMD) for the herniation of the cerebellar tonsils was Cornelis Joachimus van Houweninge Graftdijk from the Netherlands [1]. He connected the physiopathology of the cerebellar herniation with elevated intracranial pressure. He sought to increase the space through which the cerebellum had herniated with the purpose of allowing a better flow of the cerebrospinal fluid (CSF). In 1930, he operated a patient with myelomeningocele, a rapid growth of the head and a ventriculogram showing an encephalon hernia. With this surgical action he tried to alleviate the CSF obstruction due to the extruded cerebellar tissue in the cranio-cervical junction. This contribution was published in the doctoral thesis with the title “Over Hydrocephalus” [1].

The idea that the displacement of the drainage holes of the forth ventricle towards the upper end of the spinal canal could prompt hydrocephalus and that the entire anomaly could act as a valve was first expressed by Van Houweninge Graftdijk in 1932 [1]. For this case, parts of the occipital bone and posterior elements of the first two vertebrae were removed. The patient developed a fever on the second post-operative day and died 84 days after the surgery, the bladder ruptured so that the patient died on day 98 of the post-operative period.

This is the first known intent to surgically correct a posterior encephalon herniation. Van Houweninge Graftdijk also proposed the theory of the caudal traction due to the myelomeningocele as being responsible for “pulling” caudally on the posterior part of the encephalon, causing the Arnold-Chiari Syndrome Type II (ACHSII) [2]. He so established an etiological duality with a hydrodynamic cause for ACHSI and a mechanical cause due to traction for ACHSII, as it is currently for most authors.

In most publications it is nevertheless Penfield and Coburn of Montreal who appear as the first ones to apply the FMD in 1938 [3] when treating a herniation of the cerebellar tonsils in the case of a 29-year-old woman with thoracic “spina bifida” antecedents in her childhood. She presented recent hearing loss and paresis of the right half of the face. Upon physical examination the following was observed: nystagmus, absence of the right corneal reflex, trunk ataxia and diminished reflexes. The patient underwent an exploratory craniectomy of the posterior fossa with the diagnosis of a possible bilateral tumour of the acoustic nerve; she never recovered consciousness after the operation and died two months later. An Arnold-Chiari Syndrome Type II and hydrocephalus were observed in the autopsy. Penfield and Coburn suggested to leave the descended cerebellar tonsils intact in the future and to eliminate the posterior edge of the foramen magnum with the posterior elements of C1 and C2 [3].

Also in 1938, McConnel and Parker [4] published their results of posterior fossa decompression for Arnold-Chiari Syndrome Type I in five patients, with good results in two cases. In 1945, Bucy and Lichtenstein [5] informed of a successful decompression of an Arnold-Chiari Syndrome Type in a 40 year old woman without hydrocephalus. In 1948, Chorobski and Stepien [6] operated a woman with Arnold-Chiari Syndrome Type with a full resolution of her symptoms.

The current theory and practice of FMD applied to ACHSI and ISM

The most cited publication to date in relation to the treatment of the ACHSI/ISM complex is Gardner and Goodall's publication form 1957 [7] in which they treated idiopathic syringomyelia by means of a decompression of the posterior cranial fossa and sealing the hypothetic communication between the syringomyelic cavity and the forth ventricle in 17 patients; 13 improved, the syringomyelic cavity decreased in three, and there was one death.

The opening of the foramen magnum in all its variants, suboccipital craniectomy or craniotomy (SOC), with or without homologous, heterologous or artificial duraplasty in ACHSI, together with the shunting of the cavity for ISM, frequently associated conditions, are considered to be the current surgical treatments.

For some specialists, the FMD improves the permeability of the CSF circulation tracts that are supposedly compromised, even though this has not been proven at any point, but rather refuted, given that ACHSI has been observed in the embryo, prior to the formation of the cerebrospinal fluid and in the adult it can be observed in many cases on MRI, CT imaging, or through ultrasound during surgery, that there is an excellent communication between the cranio-cervical spaces.

For others, the FMD eliminates the mechanical cause of ACHSI and of some ISM: a supposed stenosis or decrease of the size of the posterior cranial fossa, due to which the cerebellum slides through the foramen magnum towards the spinal canal in form of a descent of the cerebellar tonsils, having no choice but to descent into spinal canal because of the conflict of space.

Since the posterior cranial fossa, where the cerebellum is located, has two openings, one caudally and inferior, the foramen magnum and the other cranial or superior or mesencephalic or tentorial of Pacchioni, and due to the same described mechanical conflict, the cerebellum could herniate upwards through the superior opening toward the cranial fossa, but there is no ACHSI/ISM case published with an upwards or even double herniation.

When there is a cerebellum displacement in ACHSI, it is always towards the occipital or caudal hole, additionally, in most cases an increase in the supracerebellar space can be observed. This is the arachnoid space between the superior side of the cerebellum and the inferior side of the tentorium with an overall displacement of the cerebellum towards the foramen magnum, signalling the caudal direction of the sole interceding mechanical force. Therefore, it can be ruled out that the small or stenotic posterior cranial fossa is the cause for ACHSI [9].

It is true that in many ACHSI cases there is a compromise of the volume between the bony edge of the foramen magnum and the nervous content – brain stem and inferior part of the cerebellum- and that its traumatic manipulation during the FMD can contribute to the mortality and morbidity rates of the surgical

FMD technique.

The tonsillectomy or cerebellar tonsil exeresis proposed by some authors probably adds to the mortality and morbidity rates. It can be an unnecessary and maiming surgical manoeuvre [3] for ACHSI, given that no benefit is obtained and it involves an amputation of a part of the cerebellum that can only contribute to sequelae, situations of permanent instability and vertigo [10].

Complications and Sequelae of the FMD applied to ACHSI and ISM

According to the book "The Chiari Malformations" (2013) by Shane Tubbs and Jerry Oakes [11], a complication rate of 28% is observed with the application of the FMD for ACHSI, most of them severe, such as:

- Neurological deficit: hemiparesis 0.5-2.1%, visual field alteration 0.2-1.4%, speech alteration 0.4-1%, sensory deficit 0.3 -1%, instability 10-30%.
- Cerebrospinal fluid leak 3-14%, which in turn can be a vehicle for deep infections, such as meningitis or encephalitis.
- Post-surgical intracranial haemorrhage 0.1-5% in the surgical site or in the epidural or intraparenchymatous space, causing a *de novo* neurological deficit or a worsening of the pre-existing deficit.
- Infarct-oedema up to 5% depending on the process that is causing it and its localization.
- Superficial or deep infection of 0.1 to 6.8%, that can be encephalitis, a cerebral abscess or meningitis.
- Others: hemodynamic alteration due to the manipulation of the injury or of the brain stem. Air embolism (patient in sitting surgical position). Post-surgical hydrocephalus. Pneumoencephalon.

Mortality of the Foramen Magnum Decompression applied to ACHSI and ISM

In the book "Syringomyelia - Diagnosis and Treatment" (2002) by Jörg Klekamp and Madjid Samii [12], the authors refer to their own mortality rate of 1% with regards to the FMD applied to ACHSI and ISM. Later, in 2012, Klekamp publishes a mortality rate of 1.3% with 21.8% of relevant complications.

The same book quotes the mortality rate of FMD applied in ACHSI and ISM of other authors such as Aghakhani J N, Parker F, Tadie M (1999) [13] of 0.7%, for Paul KS, Lye RH, Strang FA, Dutton J (1983) [14], it is of 1.4%, and for Di Lorenzo N, Fortuna A, Guidetti B (1982), it is of 12,1% [15].

In a bibliographical review of some of the most relevant publications where mainly the FMD was applied for ACHSI and ISM since 1982 to 2016, we can observe that the studies with few cases predominate;

most can be counted in dozens, others in hundreds and in four pieces there are more than 200 cases (table 1) [13,14, 16-34]. One of those, by Arnautevic et al (2015), is a bibliographical review of numerous series reaching 8605 cases of ACHSI [17].

Table 1.- Bibliographical summary concerned with ACHSI from 1982 to 2016

Author	N	Technique	Improved %	Complications %	Mortality %
Zuev, 2016 [16]	125	FMD	56	4.4	0
Arnautovic '15 [17]	8605*	FMD	78	4.5	3
Deng, 2015 [18]	152	sFMD	83	-	0
Kennedy, 2015 [19]	156	FMD	91	2.6	0
Chavez, 2014 [20]	177	FMD	95	-	-
Medkour, 2014 [21]	42	FMD	84	15	0
Chotai, 2014 [22]	30	wFMD	90	13	-
Gurbuz, 2014 [23]	39	FMD	70	17	-
Lee, 2014 [24]	65	FMD	15	10	-
Lee, 2014 [25]	56	FMD	92	-	-
Batzdorf, 2013 [26]	177	FMD	88	14	-
Deng, 2013 [27]	38	FMD	87	0	0
Gonçalves da Silva, 2013 [28]	192	FMD	74	-	7
Isik, 2013 [29]	44	FMD/sh	89	21	0
Alfieri y Pinna, 2012 [30]	105	FMD	90	11	0
Klekamp, 2012 [31]	359	FMD	74	22	1.3
Saceda-Gutierrez, 2011 [32]	36	FMD	84	33	0
Tubbs,2011 [33]	500	FMD	83	2.4	0
Taricco y Melo, 2008 [34]	29	FMD/sh	72	-	0.1
Aghakani, '99 [13]	285	FMD/sh	87/71	-	1.4
Di Lorenzo,'82 [14]	47	FMD	52	-	21

sFDA = small FDA; wFDA = wide FDA; FDA/sh = FDA/shunt

Due to its important case number, we consider the publication by Arnautovic et al to have a certain statistical value. The authors reviewed the publications in English of paediatric, adult and combined (paediatric and adult) series of patients with ACHSI from 1965 to 2013 and identified 145 surgical series of patients with ACHSI, mainly in the United States of America and Europe. They collected 8605 cases that were operated mostly with FMD with an overall figure of 4,5% complications and 3% mortality (average of the sixteen series that contributed mortality caseload). Extrapolated to the total, this would represent some 258 dead patients with regards to the surgical technique, being mainly FDM. Whereas the

spontaneous mortality rate of the condition ACHSI is 0.0004%, which in Arnautovic's caseload would amount to a possible 0.034 dead patient, if no patient would have undergone surgery.

Comparison between the natural course and the application of FMD for the ACHSI/ISM complex

In the last three decades, eight deaths related to the spontaneous progression of the ACHSI/ISM complex have been published. If the 2.25 million cases affected by the ACHSI/SM complex in three decades had undergone surgery with a mortality index of 3%, this would mean 7500 iatrogenic deaths as opposed to a very low spontaneous mortality.

Conclusion

Currently and thanks to the collection of series of operated patients, we know that the FMD in its indication for ACHSI and ISM involves a mortality rate of 3%. As it is justified to assume it in fatal pathologies such as tumours, hematomas, cysts and others with a high mortality rate that in many cases can be of a 100%, it is not justified in ACHSI and the ISM with a low spontaneous mortality rate.

The application of the usual means to treat ACHSI and ISM, the FMD, exceeds and adds by 3% to the spontaneous mortality rate of 0.0004 in ACHSI and 0% in ISM.

Surgical teams would need to consider in the FMD applied to the ACHSI/ISM complex, that the surgical mortality rate should be less than one death in 25 000 cases (0.0004%) for it to be equivalent to the spontaneous mortality rate and its application not to be contraindicated.

And considering the parameters of Arnautovic's publication (17), we could venture into believing that no more than 10 000 cases of application of the FMD to the ACHSI/ISM complex have been published from 1930 to today. 15 000 more cases without any fatality would be needed for the FMD's iatrogenic mortality rate to be lower than the spontaneous mortality of the ACHSI/ISM complex. These criteria are of concern in order to assess other possible treatments and surgical indications for the ASCHI/ISM complex.

The presumed improvement in some cases with the FMD applied to ACHSI and ISM does not justify the death of even a few patients.

It can therefore be deduced that the FMD with iatrogenic mortality rates that go from 0 to 12.1%, with an average of 3%, and a morbidity ranging from 3.4 to 28% with severe complications, is not indicated or rather contraindicated as the standard treatment for ACHSI and ISM. Nevertheless, the application of the FMD can be considered for ACHSI when inaction represents a greater danger than the FMD.

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