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Developmental and surgical anatomy of pathologies of the craniovertebral junction: a much-needed problematization of efficacy and long-term outcomes

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Abstract. Introduction. Due to its complex anatomy and function the craniovertebral junction (CVJ) is prone to several pathological conditions. The unique topography of neuro-vascular and bony structures makes surgical management of this area particularly challenging. Methods. A systematic online literature search was conducted in *PubMed, Google Scholar, Scopus, and Web of Science* with keywords relating to Chiari malformation, Wallenberg syndrome, Syringomyelia, and atlantoaxial dislocations. Results. This paper presents an up-to-date summary of the pathogenesis and surgical management of Chiari malformation, Wallenberg syndrome, syringomyelia, and atlantoaxial dislocation with particular emphasis on developmental and surgical anatomy. Conclusion. In such complex pathologies of the CVJ, the many factors contributing to the patient's condition and the aspects playing a key role in the treatment process are all difficult to be considered concurrently. A better and universal surgical technique is impossible to be established.

Keywords: craniovertebral junction, Chiari malformation, Wallenberg syndrome, syringomyelia, atlantoaxial dislocations, surgical management.

INTRODUCTION

The human head contributes to approximately 6% of total body weight. Therefore, an average 80 kg male individual carries five kilograms of weight at the top of his torso throughout his lifetime (Szczygieł et al., 2020). The cranium transitions through the craniovertebral junction into the spine. Several structures contribute to the junction including 6 joints in total: the upper joint of the head comprises two atlantooccipital joints, while the lower joint of the head contains four joints: two lateral atlantoaxial joints and two median atlantoaxial joints anterior and posterior (Gworys, 2012, Standring, 2021) these joints are supported by muscles, ligaments, and membranes which serve two functions: allowing movement in the upper cervical region and protecting vital structures, such as the spinal cord, nerves and vessels supplying the cerebrum. Both congenital and acquired diseases develop in this area. The complexity of the region and its anatomical variability among individuals provide challenges to surgical management.

The multistage diagnostic process usually begins with computer tomography (CT scan imaging), which has questionable accuracy and poor image resolution (Takami et al., 2014; Chi et al.,2019). In contrast, magnetic resonance imaging (MRI) and its parameters provide physicians with a more precise image of a patient's current stage. A further advantage of MRI is its ability to depict soft tissue (Winklhofer et al., 2014). A more invasive diagnostic examination is a myelogram (Kular & Cascella, 2022). This paper focuses on the pathogenesis of diseases of the craniovertebral junction and provides a comparison of specific procedures which were performed during each pathological study.

METHODS

This review was based on an internet search of *PubMed*, *Google Scholar*, *Scopus*, and *Web of Science* in order to find pathological cases based on a specific word search. Word search entries included 'Chiari malformation', 'Chiari malformation types', 'Wallenberg syndrome', 'lateral medullary syndrome', 'posterior inferior cerebellar artery syndrome', 'Syringomyelia', and 'atlantoaxial dislocations'.

RESULTS

Etiology and classification

Chiari malformation (CM, previously known as Arnold-Chiari malformation) was first observed and described by Julius Arnold (1835-1915), a German pathologist, along with his Austrian colleague Hans Chiari (1851-1916). They discovered a general deformation of the hindbrain accompanied by ectopic cerebellar tonsils. Currently, CM is classified according to four types (Table 1). Type 1 is the most prevalent type of CM, which involves a caudal displacement of the cerebellar tonsils through the foramen magnum. The displacement must be 5mm or greater with no involvement of spinal herniation to be classified as type 1 CM. Nevertheless, a spinal herniation may also appear in other types of CM. Another common characteristic of type 1 CM is its late appearance in adolescents, whereas type 2 CM can be diagnosed much earlier. Furthermore, type 2 CM is typically diagnosed with ectopy of the cerebellar vermis, fourth ventricle as well as brainstem, and myelodysplasia. Type 3 and 4 CM are extremely rare (Kular & Cascella, 2022; Fric et al., 2019; Brito et al., 2019; Milhorat, 2009). Type 0/0,5 CM has recently been identified as having no symptoms of type 1 CM in children, however, in this type, the cerebellar tonsils wrapped around the medulla oblongata. Type 1.5 combines symptoms of type 1 and 2. Additionally, there is no evident spine herniation in type 1.5 CM (Fric et al., 2019; Morgenstern et al., 2020).

No specific or supported mechanism of the pathogenesis in CM has been supported by research. Some arguments purport that CM is a genetic disease with association with alleles located on the 9th and 15th chromosomes (Boyles et al., 2006). Alternatively, counterarguments highlight that CM is rarely identified in families and that late diagnosis in infants may be asymptomatic (Fric et al., 2019). Although type 2 and 3 CM derive from neuro-ectodermal defects, type 1 CM is believed to be an effect of a para-axial mesoderm lesion leading to impaired development of the posterior cranial fossa. Interestingly enough, type 1 CM can occur alongside cases in which no cranial hypoplasia is observed, such as tethered cord syndrome, hydrocephalus, or acromegaly (Milhorat, 2009). Thus, CM can be diagnosed as a secondary condition to other diseases. Iatrogenic CM has also been reported (Kular & Cascella, 2022).

Table 1. Epidemiology of the craniovertebral junction pathologies.

Disease		Epidemiology
Chiari malformation	Type 1	1 per 1280 live births (Capra et al., 2019)
	Type 2	1 per 1000 live births (Kuhn and Emmady, 2022)
	Type 3	51 cases reported (Elbaroody et al., 2021)
	Type 4	no data available
Wallenberg syndrome		60 000 cases a year only in the United States (Venti, 2012)
Syringomyelia		8.4 per 100000 live births (Sharma et al., 2006)

Wallenberg Syndrome (WS) otherwise known as the lateral medullary syndrome was first mentioned in 1808 by Gaspard Vieusseux (1746-1814), with a more precise description of WS being presented by the German neurologist Adolf Wallenberg (1862-1949) in 1895 (Wallenberg, 1895). Vieusseux described WS as an occlusion of the vertebral artery or the posterior inferior cerebellar artery (PICA) followed by infarction of the lateral medulla oblongata (Lui et al., 2022). Lateral medullary syndrome (LMS) is the non-eponym designation of WS. In WS the lateral region of the medulla oblongata, posterior to the olivary nucleus is affected.

In order to understand the mechanism of WS a brief description of PICA is necessary. The posterior inferior cerebellar artery is a branch of the vertebral artery, arising 1,6-1,7 cm below the vertebrobasilar junction. The main trunk of PICA is the source of branches supplying the lower cerebellar hemispheres, the vermis, the fourth ventricle, and the tonsils. As mentioned, blood clots can develop in PICA or even in the vertebral artery. Moreover, PICA is important when aneurysms are involved since they are prone to rupture during surgical procedures (Lui et al., 2022). Risk factors of WS include hypertension, smoking, diabetes, or arteriosclerosis. Also, head trauma may contribute to the evolution of WS, as well as Marfan syndrome, Wegener's granulomatosis, or during a cocaine overdose (Lui et al., 2022; Gasca-González et al., 2020)

Syringomyelia which is the subsequent pathology in focus, is a condition in which a cavity filled with fluid in the parenchyma of the spinal cord occurs (Wichmann et al., 2020). It was originally described by Brunner et. al. in 1688 (Hart, 2014). The fluid-filled cavity is called the syrinx. A vital aspect of the fluid filling the cavity is that it is isotonic or close to isotonic with the CSF but with a much lower protein content (Rusbridge et al., 2006). That fluid-filled cavities must be separated from protein-filled tumor cavities which may resemble the syrinx (Rusbridge et al., 2006). Specialists describe the syrinx as a secondary lesion to another aforementioned disease of CM or myelomeningocele but with higher prevalence as it is associated with high-energy trauma (Wichmann et al., 2020; Fischbein et al., 1999). Syringomyelia can also develop in patients with no history of trauma due to cervical spondylostenosis or arachnoiditis.

Although an accurate mechanism remains unclear, the consensus is that it is caused by impaired cerebrospinal fluid (CSF) flow and partial obstruction. The theory of syringomyelia proposes that it occurs due to hypertension in the arachnoid matter the CSF is pumped through perivascular spaces into the parenchyma and accumulates in the central canal of the spinal cord. If the CSF remains in the parenchyme the condition is called presyrinx (Fischbein et al., 1999). An alternate theory argues that a cascade of this kind would rather cause increased pressure on the spinal cord; second, the differential pressure between the spinal cord and the subarachnoid space provides the driving force for such action. However, these theories do not explain the lowered protein level in the syrinx. Yet another hypothesis is described by Clare Rusbridge et al. to explore the relationship between syringomyelia and the principles of thermodynamics, specifically investigating fluid dynamics in the context of CM. The authors delve into the potential implications of fluid flow and energy considerations on the development and progression of syringomyelia, providing valuable insights into the underlying mechanisms of this neurological condition (Rusbridge et al., 2006). The authors suggested a division between communicating and non-communicating types of syringomyelia in reference to the connection between the syrinx and the fourth ventricle. The first type is diagnosed in less than 10% of patients and has received little theoretical attention (Rusbridge et al., 2006). However, some authors have speculated that the first type of syringomyelia may run in families (Busis & Hochberg, 1985).

Previous research has highlighted the vital connection between the cervical vertebrae C1 and C2 vertebrae in relation to mobility as well as instability. The principal joints of this region the atlantooccipital joint and atlantoaxial joints have various functions. While the atlantooccipital joint provides a positioning function, the atlantoaxial joint performs head rotation. That unfortunately makes it highly prone to any defects and damages. Some researchers consider that the atlantooccipital joint is a highly vulnerable structure (Goel, 2019; Salunke, 2018; Ibrahim et al., 2021).

Even small forces like a tap on the head in early childhood may lead to instabilities and minimal deformation of the upper cervical region (Goel, 2019). Instability and dislocations are frequently diagnosed in elderly people with weakened muscle tonus and children due to their smooth and slippery articular cartilages.

These conditions may contribute to the loss of correct stabilization (Goel, 2019). It has also been observed that any inborn asymmetry or malformation of the lateral masses of the atlas will lead to increased forces on one side of the joint (Salunke, 2018). Charles Bell in 1830 described a case in which a patient developed spinal cord compression secondary to pharyngitis and syphilis (Grisel, 1951). After two more cases of nontraumatic atlantoaxial dislocations (AAD), it was defined as Grisel syndrome. The mechanism of this condition remains unknown (Barcelos et al., 2014). Atlantoaxial dislocations are a frequent outcome of high-energy trauma with a high mortality rate. Odontoid fracturing often occurs in such situations (Sánchez-Ortega et al., 2021). Other risk factors include rheumatoid arthritis, tuberculosis, Hirayama disease, or myelopathy which is related to the ossification of the posterior longitudinal ligament (Goel, 2021). Some researchers claim that atlantoaxial instability itself can cause basilar invagination or the CM (Wagner et al., 2020). Two classifications of atlantoaxial dislocations have been developed. The first classification focuses on the direction and surface of the malformation therefore we have anterio-posterior dislocation, rotary, central, and mixed one. The second one is clinically divided by surgeons into reducible (RAAD) and irreducible (IAAD) atlantoaxial dislocations.

Symptoms

Since pathologies occurring in the craniovertebral junction show similarity, they are difficult to distinguish.

Symptoms of CM derive from impaired CSF flow through a very constricted foramen magnum during any neck maneuvers. Pain accompanies coughing and sneezing. Suboccipital pain radiates to the frontal and parietal areas. Other symptoms include nausea and cognitive disorders. Intracranial pressure can cause disorders of the autonomic nervous system manifested as loss of consciousness, bradycardia, tinnitus, or swallowing problems. Children with CM suffer from sleep apnea and difficulties in food intake due to CNIX and CNX dysfunction (Kular & Cascella, 2022; Fric et al., 2019).

Nystagmus, dizziness, dysphagia, ataxia, and Horner's syndrome occurring on the side of the lesion, as well as contralateral loss of pain and temperature of the body and ipsilateral loss of pain and temperature to the face are signature symptoms of WS. Another noticeable symptom of WS is hiccupping (Lui et al., 2022; (16) (17). The progression of WS can be both rapid and gradual. In both cases the symptoms are similar.

Syringomyelia is a disease that can be symptomatic and asymptomatic. Due to the individual location of the cysts, the symptoms will vary in intensity and area of radiation (Wichmann et al., 2020). Generally, patients experience pain, oversensitivity, impaired muscle function, and spasticity (Schurch et al., 1996). Some patients report loss of temperature and nociception but with intact proprioception and sense of vibration (Wichmann et al., 2020). Muscular dysfunction of one limb or neurogenic bladder is also spotted in patients with syringomyelia (La Haye & Batzdorf, 1988). Interestingly, the time between trauma and the appearance of symptoms varies from 2 months to even 30 years (Schurch et al., 1996). Atlantoaxial dislocations, especially in children tend to be asymptomatic. This unfortunately delays diagnosis and treatment. No specific neurological symptoms have been assigned to this pathology, but a cock-robin position of the head can be an indication. In this position, the head inclines to one side and tilts to the other side (Goel, 2019; Štulík et al., 2022). The literature also mentions acute neck pain and numbness of the limbs (Ibrahim et al., 2021). Dislocations occurring after highimpact trauma may cause breathing difficulties (Koljonen & Cheung, 2021; Liu et al., 2022). It is difficult for patients in the cock-robin position to turn their heads contralaterally.

Surgical management

As in most spinal defects, a patient's individual anatomy and general condition must be taken into consideration while clinical treatment of CM is being established. Though surgical management is usually a method of choice, some ideas of conservative treatment also occur in literature. This depends on whether the symptoms exacerbate, or the disease is prone to development. The Pediatric Section of the American Association of Neurological Surgeons stated that there is no need for surgical decompression procedures in asymptomatic children (Siasios et al., 2012). Conservative treatment includes non-steroidal anti-inflammatory drugs and muscle relaxants which provide minimal results in patients with more intensified symptoms.

Surgical procedures aim to re-establish equal pressure between the intracranial subarachnoid space and the intramedullary area. The cerebral spinal fluid (CSF) should have normal pressure. The main procedure performed in almost 99% of cases was the posterior fossa decompression (PFD) which focuses on widening the cisterna magna with one or two posterior arches of the cervical vertebrae and opening the dura matter but leaving the cerebellar tonsils in herniation (Fric et al., 2019; Arnautovic et al., 2015; Siasios et al., 2012; Lou et al., 2019). However, this is not a flawless procedure since the side effects may include cephalomeningitis or CSF leakage. Moreover, 17% of patients require repeated surgery (Lou et al., 2019). To decrease the side effects of PFD, a posterior fossa reconstruction (PFR) with duroplasty can be performed in type 1 CM with syringomyelia. An advantage of PFR is that it does not interfere with the CSF flow due to the remained continuity of the arachnoid matter. Currently, there is no consensus on a 'better' method for PFR. Advantages of PFD include shorter operating times and lower hospitalization costs. Research carried out on 582 patients provides information about

similar outcomes of PFD with or without duroplasty (Lou et al., 2019; Mohanty, 2019). A less invasive technique does not involve craniectomy removal of subdural tonsil herniation with a reconstruction of cisterna magna in patients with type 1 CM with syringomyelia. However, some side effects were also observed (Kular & Cascella, 2022; Lou et al., 2019). If the CSF flow does not return to its proper level after any of those procedures or the intracranial pressure increases, then a ventriculoperitoneal shunt is recommended (Fric et al., 2019).

Regarding WS, diagnostic tools are worth mentioning. MRI with diffusion-weighted imaging (DWI) is a prescribed method since it allows early detection of ischemic lesions (Lui et al., 2022; Heinemann et al., 2009). Rare cases like vertebral artery dissection can be dismissed due to the use of a CT or MRI angiogram followed by a precise location of the damaged area (Lui et al., 2022; Saleem & Das, 2022). The best-case prognosis is where a quick diagnosis is established. Some research mentions similar steps, beginning with an intravenous (IV) thrombolysis with IV tissue plasminogen activator (Lui et al., 2022; Saleem & Das, 2022). Endovascular revascularization performed with modern devices is recommended if large intracranial vessels have been impaired. Patients who have undergone this procedure tend to have improved recovery (Lui et al., 2022). Secondary prevention includes statins, antiplatelets (i.e., aspirin and antihypertensive drugs).

Using phase-contrast MRI to measure blood flow in large vessels is considered an optimal diagnostic method in syringomyelia. A benefit of this method is that it is free from side effects and provides physicians with qualitative and quantitative data. Moreover, it is less invasive than lumbar puncture or neuroendoscopy (Wichmann et al., 2020; Fischbein et al., 1999).

Surgical treatment is performed in symptomatic cases in which patients' mobility is impaired and limited or where there is inordinate pain. Conservative treatment is sparsely used in symptomatic cases (Schurch et al., 1996). An interesting position among surgical maneuvers is the syringo-subarachnoid-peritoneal shunt with a T-tube. According to some authors, this method differs from syringe-subarachnoidal or syringe-pleural shunts which are also used to siphon off CSF. Unfortunately, a drawback of syringe-subarachnoidal or syringe-pleural shunts is their tendency to fail. In the case of syringepleural shunts repeat surgery may be required within a year (Kim et al., 2012). Other disadvantages of shunts are their incidence of displacement and obstruction. For example, a linear catheter in syringostomia runs a high risk of dislocation. The method presented in the article included high perforation of the T-tube and allocation of only one arm of the tube in the syrinx and the second in the subarachnoid space to avoid injuring the spinal cord during its removal. In one patient who received this treatment, his syrinx decreased with subsequent improvement in motor function in his upper right arm (Kim et al., 2012).

Other articles which mention the T-tube as a shunt of choice do not evaluate any specifics of the procedure. Syringostomia tends to have a positive outcome in patients most of whom report decreased symptoms (Schurch et al., 1996). In one of the positions available after individual assessment doctors performed: suboccipital decompression with partial resection of cerebellar tonsils or C3 through C7 laminectomy with sectioning the dentate ligament at C3-C4 and lysis of subdural adhesions or VP shunting. Even though some patients had suffered from other disorders aside from syringomyelia, in most of them, an improvement was observed (Fischbein et al., 1999). Another research used cyst-toperitoneal shunt in most patients all of whom regained physical strength and their pain decreased (La Haye & Batzdorf, 1988).

In Goel and Desai's research, an interesting aspect was that a syringe-subarachnoidal shunt was used to treat syringomyelia secondary to CM and basilar invagination (Goel & Desai, 2000). Here, the shunt was a fundamental procedure. Although the operation may succeed in decreasing the size of the syrinx, symptoms caused by the pressure on the brainstem and cervical spinal cord may increase. The authors point out that the syrinx might have evolved as a protective phenomenon to protect vital structures prone to any damage because of the CM and not as a pathological condition (Goel, 2001).

Between 2002-2015, a series of surgical procedures containing laminectomy, lysis of adhesions, cyst fenestration, and duraplasty was performed in patients with post-traumatic syringomyelia. The outcome was ambiguous since some patients reported improvement and some claimed that the symptoms declined (Li et al., 2021). Patients often required a repeated operation after surgical management in syringomyelia, in some cases there is a need for even 10 re-operations when spine fusion is performed. Circumferential spine surgery, lysis of adhesions, or duraplasty are believed to be the most effective in decreasing the risk of repeated operation (Li et al., 2021).

Before proceeding with the surgical methods in atlantoaxial dislocations, some discussion on the aims of treatment is necessary. The inclination should be reduced with traction (closed method) and if not possible by opening the joints (open method). No pressure on any vital structures should remain post-operation. Usually, an arthrodesis is recommended (Jain, 2012). In Among techniques used in RAAD treatment after the repositioning of the joint and bone graft, is sub-laminar wiring. In this procedure, complications may occur involving cutting the posterior arch of the atlas with the wire. Titanium is often used due to its pliability when compared to steel rods. It should be pointed out that a fusion from the occiput to C4 should be performed only when a patient requires it. Another disadvantage of such fusion from the occiput to C4 is the high force impacting the structures at both ends. The C1-C2 spacer jamming is a further option but should be executed along with sub-laminar wiring or C1-C2 fusion to strengthen the connection. A trans-articular screw with sub-laminar wiring by the Magerl technique is also considered functional, stable, and efficient (Jain, 2012).

Atul Goel proposes that the first approach to reducing a dislocation can be cervical halter or halo-gravity traction. If after 3-4 weeks no improvement is observed, an open procedure can be implemented. The Goel method allows the patient to have greater freedom of movement in the cervical region (Goel, 2019). This method has proven to be successful. Prevalent procedures are C1-C2 posterior fixations executed by transarticular Magerl or the above-mentioned Goel technique. New solutions include transpendicular axis mass fixation and an anterior transarticular fixation with the Vaccaro method (Sánchez-Ortega et al., 2021).

Not many papers mention the management of atlantoaxial instabilities in the Grisel syndrome. In this case, conservative treatment precedes surgical intervention. A protocol of conservative treatment in atlantoaxial dislocations is mentioned by Wetzel and La Rocca who discuss the management of dislocations emerging in children in relation to a 7-year-old male patient. Due to the patient's age and body build, a fusion would have had a high risk considering the size of the child's bones and soft tissues. The authors mention Menezes who proposes, in this case, a subtle craniectomy with laminectomy of the upper cervical region and a bilateral interlaminar followed by occipital rib graft. This is fixed with a titanium wire (Barcelos et al., 2014).

Harms-Goel and Magerl techniques are mentioned along with the Wright method in which the screw is allocated through the lateral masses of the atlas and intralaminarly of the axis (Marcon et al., 2012). In the case of IAAD, a transoral odontoidectomy is performed, although some surgeons prefer a more applicable method of transoral decompression. A transoral odontoidectomy is aimed at delivering the neurological structures (Jain, 2012). This procedure has a high risk of CSF leakage and meningitis (Liu et al., 2022). Halo traction and vest are listed again as ways for managing acute atlantoaxial dislocations in children with no guarantee of

toaxial dislocations in children with no guarantee of reduction. Surgical operations involved here are Magerl, Harms-Goel technique, and Brooks or Gallie wiring (Koljonen & Cheung, 2021).

DISCUSSION

Undoubtedly, the complexity of the cranio-vertebral junction has several drawbacks which need to be considered before surgery. Their etiology is probably an outcome of many factors including a genetic base possible in the development of CM or syringomyelia (Boyles et al., 2006; Busis & Hochberg, 1985). High-impact trauma, on the other hand, may contribute to the increase of syringomyelia or atlantoaxial dislocations (Wichmann et al., 2020; Fischbein et al., 1999; Sánchez-Ortega et al., 2021). The occurrence of one pathology in this area does not exclude others. For example, syringomyelia may be secondary to CM. Symptoms of both diseases often resemble each other.

As for the treatment, milestone steps have been made. A posterior fossa reconstruction aims at eliminating side effects of PFD in CM which is found in the literature. Nevertheless, many surgeons perform a PFD with various levels of success (Lou et al., 2019; Mohanty, 2019).

Magnetic resonance imaging with diffusion-weighted imaging (DWI) which is used in Wallenberg syndrome diagnostics allows a fast and accurate diagnosis (Lui et al., 2022; Heinemann et al., 2009). Additionally, endovascular revascularisation provides a patient with a higher rate of recovery (Lui et al., 2022).

The treatment of syringomyelia offers several tools that can be employed in the treatment process. The syringo-subarachnoid-peritoneal shunt, as the authors propose, is a questionable option since the obstruction and moveability of shunts have to be taken into consideration (Kim et al., 2012). Other surgeons perform a series of cyst-to-peritoneal shunt placements with a highly positive outcome (La Haye & Batzdorf, 1988). Laminectomy, lysis of adhesions, cyst fenestration, and duraplasty used in the treatment of patients found in one article had questionable results (Li et al., 2021). The idea that syringomyelia is an evolutionary development for decreasing pressure still requires more research (Goel, 2001).

In relation to atlantoaxial dislocation, the Goel and Magerl method in posterior fixation is often preferred (Goel, 2019; Koljonen & Cheung, 2021). Other aspects have been addressed in the medical literature, including the unnecessary occiput to C4 fusion which can limit a patient's movements. This systematic review found few papers discussing the treatment of Grisel syndrome. In relation to children's atlantoaxial dislocations, Menezes management has been examined (Barcelos et al., 2014).

CONCLUSIONS

The aim of this paper was to review the surgical aspects of cranio-vertebral junction pathologies. Current knowledge of each disease's etiology and treatments was included. Although several surgical techniques are currently in use, more research is necessary to identify any long-term effects. Fortunately, due to ongoing developments in medical technology, surgeons have an increasing number of resources to use. A conclusion drawn from this paper is that a universal technique for correcting pathologies of the craniovertebral junction is not possible. This is due to the complex nature of pathologies, as well as various other factors contributing to a patient's condition which play a vital role in the treatment process, and which are difficult to be considered concurrently.

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