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Surgical Treatment of Scoliosis with Diastematomyelia
Traitement chirurgical de la scoliose avec diastomatomyélie

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Abstract

Diastematomyelia is a congenital malformation of the spinal cord defined by the existence of two cords separated by a bone, cartilage, or fibre septum. It is associated to spinal malformations whose surgical care are difficult and risky as it may entail severe neurological complications.

The aim of this study is to evaluate the necessity, if any, of prior detethering of spinal cord before surgical correction of scoliosis.

The series of the present study comprises nine cases of congenital scoliosis with diastematomyelia.The scoliosis progress requires surgical intervention. A preoperative traction has been performed for a period of two to three weeks, followed by a minimally invasive surgery of the malformed area (diastematomyelia).

The results showed that the nine patients have been surgically treated for their scoliosis. Among the surgical techniques used is the correction of the scoliotic curve using vertebral instrumentation without prior resection of the septum.

Post-operative results showed a 70° decrease in the scoliotic curve. However, no post-operative complications, namely infectious or neurological, were noticed on these patients.

In diastematomyelia scoliosis cases, therapeutic attitudes are controversial. Some authors advocate the importance of the detethering of the spinal cord and the resection of the septum before the scoliosis surgery. For other authors, the scoliosis surgery is performed straight away. The neurological deficits are accounted for by the importance of kyphosis more than by the intra-spinal anomaly.

Keywords: scoliosis, diastematomyelia, intraspinal malformation, minimally-invasive-surgery.

Résumé

La diastomatomyélie est une malformation congénitale de la moelle définie par l'existence de deux cordons médullaires séparés par un septum osseux, cartilagineux ou fibreux. Elle s'associe à des malformations rachidiennes dont la prise en charge chirurgicale est difficile avec risque important de complications neurologiques graves.

L'objectif de ce travail est d'évaluer la nécessité ou pas de la libération préalable de la moelle avant la correction chirurgicale de la scoliose.

Notre série comporte 09 cas de scolioses congénitales avec diastomatomyélie). L'indication chirurgicale a été posée devant l'évolutivité de la scoliose. Une traction préopératoire a été réalisée pendant 2 à 3 semaines, suivie d'une chirurgie mini-invasive sans chirurgie préalable de la zone malformée (diastomatomyélie).

Les résultats ont montré que les neuf patients inclus ont été traités chirurgicalement pour leur scoliose. Les techniques chirurgicales utilisées comprenaient la correction de la courbure scoliotique par instrumentation vertébrale, sans résection préalable de l'éperon. Les résultats postopératoires ont montré une réduction de la courbure scoliotique avec un gain de l'ordre de 70°. Cependant, aucune complication postopératoire n'a été observée chez ces patients, notamment infectieuse ou neurologique.

Dans les scolioses avec diastomatomyélie, les attitudes thérapeutiques sont controversées. Certains auteurs exigent la libération de la moelle et la résection de l'éperon avant la chirurgie de la scoliose. Pour d'autres cette chirurgie première de libération de la moelle n'est pas nécessaire sauf s'il existe un déficit neurologique évolutif . Dans notre série, la chirurgie de la scoliose a été réalisée d'emblée. Les déficits neurologiques s'expliqueraient plus par l'importance de la cyphose que par l'anomalie intra-médullaire.

La chirurgie de correction de la scoliose avec diastomatomyélie reste controversée. La chirurgie de correction d'emblée sans libération préalable de la moelle est une alternative justifiée étant donné que les déficits neurologiques seraient plutôt liés à l'importance de la cyphose.

Mots clés : scoliose, diastomatomyélie, malformation médullaire, chirurgie mini invasive.

Introduction

Congenital scoliosis happens as a result to vertebra development anomalies, namely development and segmentation anomalies. It is also frequently associated with malformations of the spinal cord such as diastematomyelia. This latter is a form of rare spinal malformations among the interesting group of spinal dysraphism. They are characterised not only by the diversity of clinical and radiological presentations, but also by the controversy over its exact definition and therapeutical approach.

The double neural tubes syndrome is characterised by the division into two parts of the spinal cord. The hemicords can be in its own dural sheath (type 1 diastematomyelia) (fig.1) or in a common sheath (type 2 diastematomyelia) (fig.2). It can be isolated or associated to anomalies of vertebral bodies segmentation. It is generally associated to

malformations like myelomeningocele, meningocele, dermoid cysts, lipoma, syrinx, and short filum.

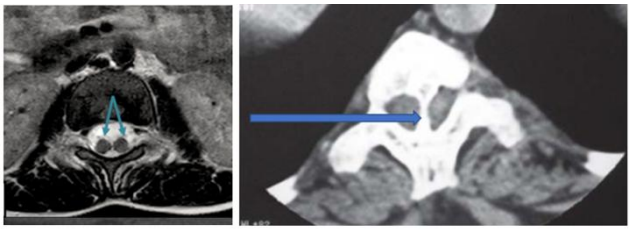

Fig.1: Diiastematomyelia type1

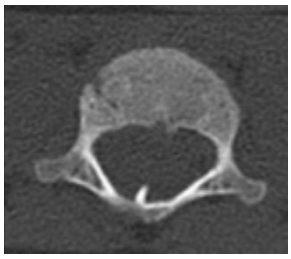

Fig.2: Diastematomyelia type2

It often associates with the vertebral malformations responsible of scoliosis and kyphoscoliosis, causing a serious problem in therapeutic care. The treatment of scoliosis associated with d diastematomyelia (fig3) raises the question of identifying optimal mechanisms to manage patients' cases while avoiding neurological complications to obtain a satisfactory correction.

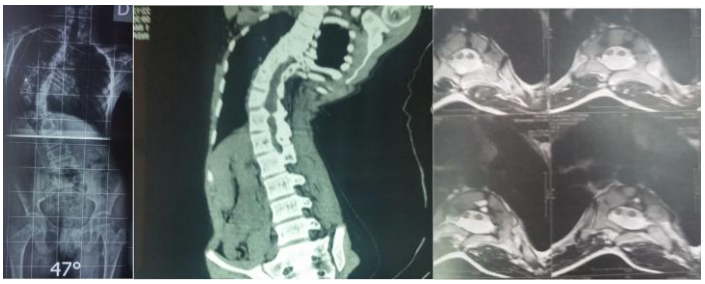

Fig.3: Scoliosis with diastematomyelia

Some authors advocate the resection of the septum before the surgical correction of the deformation as a prophylactic measure. Others perform interventions from the outset without any prior excision of the septum.

The objective of this work is to evaluate the necessity, if any, of the resection of the septum before the surgical correction of the scoliosis.

Materials and methods

Inclusion criteria are congenital scoliosis associated to diastematomyelia, an absence of neurological disorders or a neurological stability for at least the last two years. Also important to inclusion the absence of neurological complications during the traction or during radiographic dynamics as the fulcrum bending.

Cases showing neurological instability or developing neurological disorders during traction or radiographic dynamics were not included.

Our series comprised nine patients gathered between 2016 and 2019. Seven of them are girls while two are boys. All the included patients benefitted systematically from radiographs of the spine with radiographic dynamics, subtraction, along with TDM and IRM.

The localisation of diastematomyelia was thoracic among six of the patients, whereas among the three other cases it was lumbar. In all the cases, it was associated to other anomalies, namely syringomelia, butterfly vertebral malformation, segmentation defects.

The progress of scoliosis and the degree of the deformation called for surgical intervention. All the patients benefitted from traction for a duration ranging from two to three weeks.

The surgical technique consisted in a bipolar construct with a proximal fixation of laminar pedicle hook claws along with a distal fixation with two iliosacral screws. No direct access was performed in the malformed area. This latter was bridged with assembly among the nine patients.

Results

Cobb angle was found to be 71° in average with deformations that can reach 180°. The average gain of correction in the last control was 70° (fig.4). No haemorrhagic or infectious complications were noted; the neurological condition remained stable in the post-operatory control and in the follow-up.

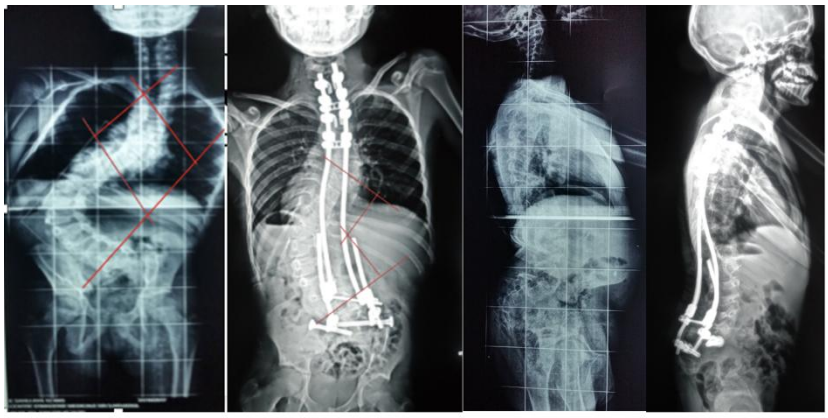

Fig.4: Correction of scoliosis with diastematomyelia

Discussion

The surgical correction of congenital scoliosis associated to diastematomyelia requires from us a careful thinking about a better therapeutical strategy in order to avoid possible irrevocable neurological complications on the one hand, and on the other hand to obtain an optimal correction.

The question raised regarding the approach that should be followed with the distematomyelic septum is whether resection is imperative or not before the surgical correction of the scoliosis.

Traditionally, for numerous authors, the resection of the bone septum is recommended **[1,2]** before any surgical correction of deformations is started. According to McMaster **[1]**, the first resection of the bone septum constitutes a prophylactic measure among children under the age of six. For Miller et al **[3]**, based on a study conducted on 33 patients, the excision of the septum did not entail any dramatic improvement in the pre-existing neurological condition. It was even noticed that neurological symptoms exacerbated among one patient. Also reported were neurological complications, dural tear and post-operative infections between 7 and 31 % **[4,5].**

Therefore, the resection of the septum may not, improve the neurological condition; moreover, it can even exacerbate it. Certain studies have shown that septum resection can be a source of complications such as scarifying the posterior bone elements, diminishing the recipient cite of the bone graft and probably being the cause of septum regeneration [**6,7**]. Accordingly, before venturing the resection of the septum, it is important to consider those risks.

Clinically, the patients remain neurologically stable, and no neurological disorders are developed after a big traction. A study conducted by surgeons in Hong Kong **[8]** has shown that tests in hyperflexion of the spine and a traction of ten seconds can predict with a precision close to 100% that the correction with instrumentation is possible if these manoeuvres do not cause neurological disorders.

The reported prevalence of neurological complications of septum resection surgery varies between 0 % and 30%, whereas in the case of the present series, the neurological complications were 0%.

The limitations of the present study can be summarised in the following:

The small sample does not allow drawing reliable conclusions. Additionally, the correction was not highly important. Also, one patient out of nine has not reached the bone maturity. A long term follow-up is, obviously, necessary to evaluate the life quality of patients and not only the correction rate.

Conclusion

Based on the findings of this work, the surgery of diastematomyelia scoliosis without resection of the septum may be riskless if the neurological condition of the patient is stable. It is safer if the patient receives a prior traction, and mainly if PES are used as monitoring aids. This plays an important role in the prevention of neurological complications.

Aggressive distraction forces must be avoided so that the spinal cord will not be pulled. Satisfaction with a correction that yields adequate coronary balance seems to be of great importance.

Declaration of links of interest

The author declares having no conflicts in interests.

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