Arthrodèse de la colonne avec ou sans fixation du bassin oblique chez l'enfant neurologique et révision de la physiopathologie et du traitement de la scoliose neuromusculaire

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ARTHRODESE DE LA COLONNE AVEC OU SANS FIXATION DU BASSIN OBLIQUE CHEZ L'ENFANT NEUROLOGIQUE ET REVISION DE LA PHYSIOPATHOLOGIE ET DU TRAITEMENT DE LA SCOLIOSE NEUROMUSCULAIRE

La Faculté de médecine, sur le préavis de Monsieur André KAELIN, professeur associé au Département de chirurgie, autorise l'impression de la présente thèse, sans prétendre par là émettre d'opinion sur les propositions qui y sont énoncées.

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Jean-Louis Carpentier
Doyen

N.B. - La thèse doit porter la déclaration précédente et remplir les conditions énumérées dans les "Informations relatives à la présentation des thèses de doctorat à l'Université de Genève".

A Marcela pour son soutien

A Camila et Eugenia

A ma mère et à la mémoire de mon père

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Résumé

Les enfants atteints d'affections neurologiques présentent un grand risque de développer une déformation spinale. Cette déformation peut aussi être liée à un bassin oblique qui peut provoquer des points d'appui ischiatiques et éventuellement des escarres.

Cette association entraîne beaucoup de difficultés pour la prise en charge chirurgicale et la question est de savoir si le bassin doit être pris dans la fixation de la colonne.

Sur 31 patients neurologiques opérés à l'hôpital Cantonal de Genève par une spondilodèse il y a eu 12% de correction du bassin oblique quand la fusion était pratiquée jusqu'à L5, et 54% quand elle était faite jusqu'à S1. Le bassin oblique résiduel est de 16,6° en moyenne et seulement 2 patients ont eu des difficultés liées à cette déformation. En conclusion, le bassin oblique jusqu'à 25 degrés est acceptable sans risque pour cette population.

Titre: Arthrodèse de la colonne avec ou sans fixation du bassin oblique chez l'enfant neurologique et révision de la physiopathologie et du traitement de la scoliose neuromusculaire.

1) Introduction en Francais:

Les enfants atteints d'une affection neurologique ont un grand risque de développer des déformations spinales (22,32,5). La courbe la plus fréquente est une longue courbe qui prend la colonne dorsale et lombaire, pouvant être liée à un bassin oblique (60%) et à une perte de la stabilité assise.

L'incidence de cette affection est très élevée (65%) chez les patients qui ne peuvent pas marcher et qui ont une atteinte psychomotrice très importante. Des séries avec des patients déambulateurs avec peu d'atteinte centrale sont liées à une incidence de scoliose neuromusculaire de seulement 5%.

Les enfants ayant une grosse atteinte neurologique doivent rester assis pendant la plupart de la journée et la scoliose provoque des difficultés dans la vie quotidienne. Cette courbe du tronc et du bassin est associée à une instabilité du tronc qui oblige le patient à utiliser ses membres supérieurs pour maintenir le contrôle de son corps, provoquant une réduction de la fonctionnalité de l'enfant.

La scoliose peut aussi être liée à un bassin oblique qui peut provoquer des points d'appui ischiatiques et éventuellement des escarres (56).

Le traitement du bassin oblique et de la colonne neurologique a toujours été un défi.

Le traitement chirurgical de ces patients reste une intervention très importante et complexe chez des enfants aussi gravement atteints au niveau psychomoteur. Des questions éthiques se posent avant chaque geste.

La première approche est l'adaptation des moyens auxiliaires pour contenir la scoliose.

On peut commencer par le moulage de la coque pour le fauteuil roulant ou par celui d'un corset en mousse pour obtenir une position plus acceptable du tronc. Le but de cette action est de rendre l'enfant le plus indépendant possible et lui permettre une socialisation avec ses proches. Si ces moyens sont efficaces, son tronc sera plus stable et l'enfant sollicitera moins ses membres supérieurs pour se maintenir assise, ce qui lui apportera plus de liberté. Par contre, il est bien démontré dans la littérature que ces appareillages - le corset ou le moulage du dos adapté - ne peuvent pas arrêter la progression de la déformation scoliotique ; la seule raison de les mettre en place est d'améliorer la posture et de permettre au patient une meilleure tenue pour mieux gérer sa vie quotidienne.

Si ces appareillages sont un échec, en ne permettant pas d'avoir une posture acceptable du tronc, ni du point de vue de l'enfant ni, la plupart du temps, de celui de ses parents, l'acte chirurgical s'impose.

Techniquement, les indications chirurgicales de l'arthrodèse de la colonne sont :

1) la difformité de la colonne qui ne permet pas à l'enfant de s'asseoir longtemps, de respirer, de manger et de rester confortable ;
2) l'incapacité de maintenir la peau intacte suite à des escarres ;
3) une courbe peu importante mais dont on peut prédire, dans l'évolution naturelle de la maladie, une progression au point que des troubles cardio-pulmonaires commencent à apparaître secondairement à cette déformation.

La contre-indication de la chirurgie est principalement l'incapacité de l'enfant à avoir un contact visuel ou tout simplement un contact avec son entourage. Cette décision reste du point vue éthique très difficile.

Les gestes chirurgicaux ont beaucoup changé durant ces vingt dernières années. On est passé de l'arthrodèse de la colonne fixée avec des plâtres à des fixations avec des tiges, des vis et des crochets de 3ème génération qui permettent la mobilisation précoce et moins de problèmes d'hygiène.

Comme on en discutera largement pendant cette présentation, la scoliose peut avoir plusieurs types de courbure, ce qui impose des stratégies chirurgicales différentes.

Ces courbures peuvent être associées à une obliquité du bassin et cette association donne beaucoup de difficultés pour la prise en charge chirurgicale. La question est de savoir si le bassin doit être pris dans la fixation de la colonne.

Il a été démontré par plusieurs auteurs que la correction de la scoliose peut se faire correctement avec une arthrodèse de la colonne en prenant ou non le bassin.

Mais le désaccord survie si le patient présente un bassin oblique, celui-ci ne pouvant être corrigé que s'il est inclus dans l'arthrodèse. La question reste toujours de savoir si on doit ou non corriger le bassin oblique.

La raison de cette controverse est que la fixation du bassin avec l'arthrodèse de la colonne implique un temps opératoire plus long et des difficultés supplémentaires du point de vue technique. Gau (21) et Allen (1) ont démontré que la fixation additionnelle de la charnière lombosacrée ajoute 10 à 15% de complications (deux cas de pseudarthrose de la jonction lombosacrée, une fracture de tige à ce même niveau et une tige mal placée dans l'ilium sur 40 patients opérés). D'autres auteurs décrivent moins de complications : Yaszici signale deux complications mineures (protrusion des tiges dans l'ilium avec la fixation type Unit Rod) sur 47 patients. Maloney, en utilisant le même système de fixation, ne signale aucune complication sur 10 patients.

Malgré cela, l'opinion dans la littérature reste partagée : Drummond (10) écrit dans son article que le bassin oblique ne doit pas être accepté parce qu'il peut produire des points de pression et des escarres au niveau ischiatique. Mubarak (39), dans la même ligne de pensée, dit que le bassin oblique de plus de 15° doit être fixé parce qu'il peut continuer à basculer dans le futur ; par contre Gaine (20) accepte des bassins obliques de 30° sans démontrer des difficultés pour le patient. Il fixe la colonne jusqu'à L5 sans prendre le sacrum, diminuant ainsi le temps opératoire.

Une deuxième controverse est le fait que le bassin non inclus dans l'arthrodèse peut évoluer en une décompensation majeure avec une obliquité qui augmente avec le temps. Sur ce point là aussi les auteurs ont des divergences d'opinion.

Dans le service d'orthopédie pédiatrique de l'Hôpital Cantonal de Genève, on a pris l'option d'accepter les bassins obliques présentant des degrés importants d'obliquité pour diminuer le temps opératoire et les complications liées à l'arthrodèse L5-S1. Le bassin oblique est réduit chirurgicalement dans deux cas : lors d'une obliquité qui suit la courbe lombaire, c'est-à-dire que le bassin est une vertèbre de plus dans la courbe lombaire, et en cas des obliquités de bassin supérieures à 20°.

Comme la plupart des chirurgiens se sont exprimés dans la littérature en s'opposant à cette décision, nous avons voulu faire une recherche rétrospective de tous les cas opérés dans notre service qui ont présenté une scoliose neurologique et qui ont bénéficié d'une fixation de la colonne jusqu'à L5 ou S1.

L'esprit de cette recherche est de

1) évaluer la correction du bassin oblique avec la fixation de l'arthrodèse de la colonne jusqu'à L5 ou S1 ;
2) évaluer la perte de correction du bassin oblique après la fixation à L5 ou S1 ;
3) considérer les complications techniques liées à ses deux niveaux différents de fixation et les complications de la vie quotidienne pour le patient qui doit accepter un bassin oblique plus marqué.

Title: The Management of Pelvic Obliquity with Spinal Fusion in Patients with Neurological Disorders and a Review of the Pathophysiology and Treatment of Neuromuscular Scoliosis

2) Introduction:

Scoliosis is a significant problem in children with neurological disease. These patients have a high risk of developing spinal deformities (22, 32, 5). The most common scoliotic curve found in this type of patients is a thoraco-lumbar C curve which may also be associated with pelvic obliquity (60%) and with a loss of postural stability, especially when sitting. The incidence of this deformity is higher in patients who are non-ambulatory and who have total body involvement (65%). Studies of ambulatory patients with milder neurological involvement yield smaller incidences of scoliosis (5%).

Scoliosis causes problems in the in sitting position, which is particularly troublesome for the wheelchair-bound child. Trunk deformity and pelvic obliquity produce postural instability that compels the patient to use his upper limbs to keep control of his body, thereby severely reducing the child's ability to perform even the most simple of tasks for himself. Pelvic obliquity is also related to pressure points at the ischium, which leads to pressure sores and ultimately scarring (56).

The management of pelvic obliquity and neuromuscular scoliosis is a clinical challenge. Spinal surgery remains the intervention of choice in more severely handicapped patients. Ethical issues arise before each intervention, as a balance has to be found between alleviating disability and not exposing the child to overly onerous surgical procedures.

The first step in the management of the spinal deformity and pelvic obliquity is with auxiliary devices to contain the deformity. This may be done with a moulded sitting support that can be attached to the wheel chair or with a soft thoraco-lumbar brace that can improve trunk position. The goal of this treatment is to allow the child to be more independent and to be able to better socialize with his friends and family. As the trunk is more stable with these accessories the patient will be able to free his upper limbs to do other things. However, it has been demonstrated in the literature that braces and moulded sitting supports don't change the natural history of the progression of the scoliosis. These treatments lead to better trunk control which in turn simplifies daily living and may slow down the progression of the curve and delay the need for surgical intervention.

When conservative treatments fail, and an acceptable trunk control can't be maintained, a surgical approach is the next option. The surgical indications are: breathing difficulties, trouble with seating, eating disturbances related to spinal deformity, pressure sores and curves where the natural history is ineluctable progression leading to cardiopulmonary complications. Surgery is contraindicated when the patient is blind and there is no contact

with the surrounding environment. Surgery has undergone considerable changes over the past thirty years. Initially, spine arthrodeses were fixed with a cast for several months, whereas nowadays they are fixed with rods, hooks and screws that allow the patient more freedom of movement after surgery, accomplish better spine reduction and facilitate hygiene, but complications still occur.

The neuromuscular spine deformities have different etiologies, with different curve patterns and different surgical strategies. Scoliosis may be associated with pelvic obliquity and the management of these two deformities is a challenge (5). The scoliotic deformity is fixed with a spine arthrodesis, which may or may not include the pelvis, but the pelvic obliquity can only be corrected if the pelvis is included in the arthrodeses. The question remains open as to whether the latter should be included in the arthrodeses. The controversy is related to the fact that when the pelvis is included in the spinal fixation surgery lasts longer with increased technical complications. Gau (21) and Allen (1) state that the fixation of the lumbosacral area leads to an increased rate of complications to 10-15% (two pseudoarthroses and one rod fracture in the lumbosacral area, one rod protrusion in the ilium in a series of forty patients). Others authors describe fewer complications, Yaszici et al (65). mention two incidences of tips protruding into the ilium out of forty seven patients. Maloney (37), in ten patients fused with unit rods, did not encounter complications in the lumbo sacral junction.

Drummond (10) states that pelvic obliquity shouldn't be accepted because it can cause pressure sores and scarring. Mubarak (39) et al argue that pelvic obliquity with more than 10 to 15° should be fixed for this same reason and because further progression can happen after fusion if the pelvis is not included. Recently, Gaine (20) has accepted pelvic obliquity up to 30°, without complications for the patients. They fix up to L5 with less surgical time and without problems with pressure sores. Nevertheless, the literature is controversial on this topic.

At the Pediatric Orthopedic Service of the Geneva University Hospital, we accept some degrees of pelvic obliquity to reduce operating time and attendant complications due to fusion of the lumbo-sacral area. The pelvic obliquity is included in the spinal fusion when it follows the lumbar curve and when the obliquity is superior to 20°. As many surgeons disagree with this approach, we decided to undertake a retrospective study of all the patients with neuromuscular scolosis and spinal fusion up to L5 or S1.

The objectives of this research are:

To evaluate the pelvic obliquity correction with fusion to L5 or S1

To assess the outcome of the pelvic obliquity when the spinal fusion has been taken up to L5 or S1

To review complications related to the two different levels of fusion

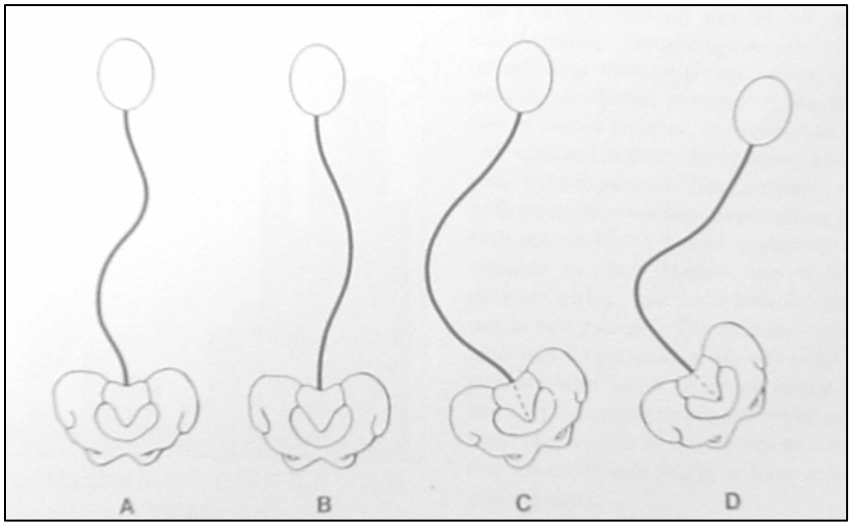
To appraise daily life impediments related to pelvic obliquity

3) Classifications

Lonstein and Alkbania classification: This classification looks for spinal deformity, balance and compensated curve as well as for the relationship with the pelvic obliquity (Fig.1). This classification is useful for our study because it combines the elements of scoliotic deformity and the pelvic obliquity. We make use of the Lonstein classification to discuss treatments and results in this paper.

- Group I: Double curves with thoracic and lumbar component. There is little pelvic obliquity and either the patient is well balanced (A) or the thoracic curve is more significant, with a fractional, poorly compensated curve below it (B).
- Group II: Lumbar or thoracolumbar curves with marked pelvic obliquity. There may be a short fractional curve between the end of the curve and the sacrum (C) or the curve may continue into the sacrum (D).

Fig 1: Lonstein classification for neuromuscular scoliosis (32)



Group I curves are most often seen in patients with mental retardation and in those who are ambulatory and live at home. These curves are associated with fewer hip contractures and fewer subluxated and dislocated hips. The average curve are not significant.

Group II entails more severe lumbar or thoracolumbar curves with marked pelvic obliquity. Sixty per cent of these patients have had a short fractional curve between the end of the curve and the sacrum, and in 40 per cent of these cases the curve continues into the sacrum. These curves occur more frequently in cerebral palsy patients with spastic quadriplegia, in non-ambulatory patients, and in patients who are not easily cared for at home. The average curve is larger, and nearly all of these patients have significant pelvic obliquity.

When the two subtypes of curve patterns are compared, the patients with a fractional lumbosacral curve show a higher incidence of mental retardation and are more likely to be ambulatory. On the other hand, the patients in whom the sacrum is part of the curve have a higher incidence of spastic quadriplegia and of dislocated hips, and their pelvic obliquity is more pronounced.

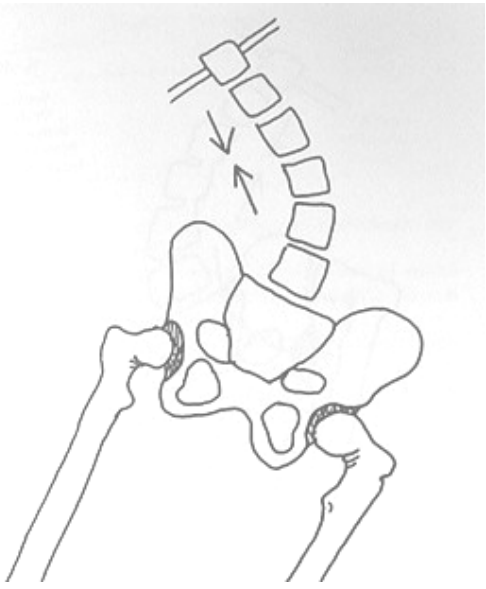
- **J. Dubousset Classification**(14) of the pelvic tilt is based on the concept that considers the sacrum and the pelvis as being one segment of the spine. He assesses the pelvic tilt in all three planes, frontal, sagittal and rotational tilt. He advocates supra-pelvic, intra-pelvic and infra-pelvic alterations as causative factors.

- Supra-pelvic factors include neurogenic disorders and congenital deformities.
- Intra-pelvic factors are deformities of the sacro-pelvic unit itself caused by trauma, infection, growth disturbance and congenital deformities.
- Infra-pelvic factors are malformations around the hip joints caused by neurogenic or other disorders.

- **Frischhut classification** (17) also applies Doubusset's concept.

  - **Type 1**: Pelvic tilt and scoliosis

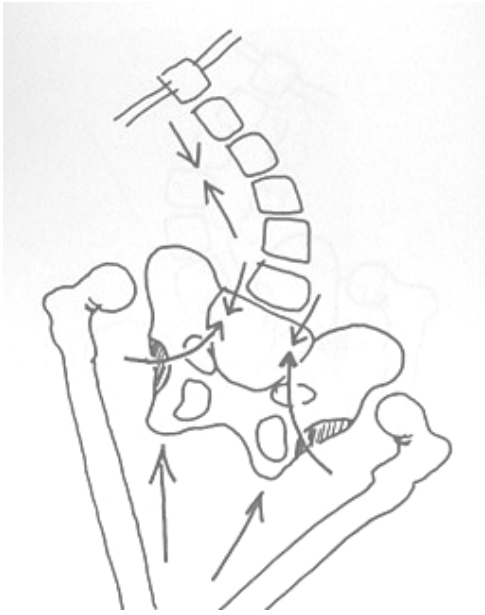
Fig 2: Frischhut classification Type 1



The deforming forces act upon the spine and the sacro-pelvic unit causing scoliosis and a pelvic tilt (Fig.2). There is no muscle imbalance around the hip causing subluxation or dislocation. This condition can be found in cerebral palsy with muscle imbalance at the trunk. In Duchenne muscular dystrophy, both muscle weakness and the forces of gravity contribute to the development of this deformity. Muscle weakness around the hip is evenly distributed and therefore hip deformities do not develop.

- **Type 2**: Pelvic Tilt, scoliosis and bilateral hip dislocation

Fig 3: Frischhut classification Type 2

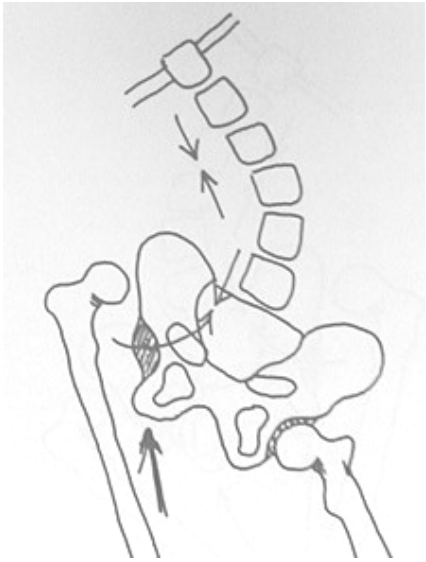


Symmetrical bilateral muscle imbalance around the hip must be viewed as a causative factor for bilateral hip dislocation (Fig.3). An asymmetrical muscle tone at the trunk induces the

development of scoliosis and a pelvic tilt. This condition is mainly found in cerebral palsy with total body involvement but also in spinal muscular atrophy.

- **Type 3**: Wind blown hip syndrome

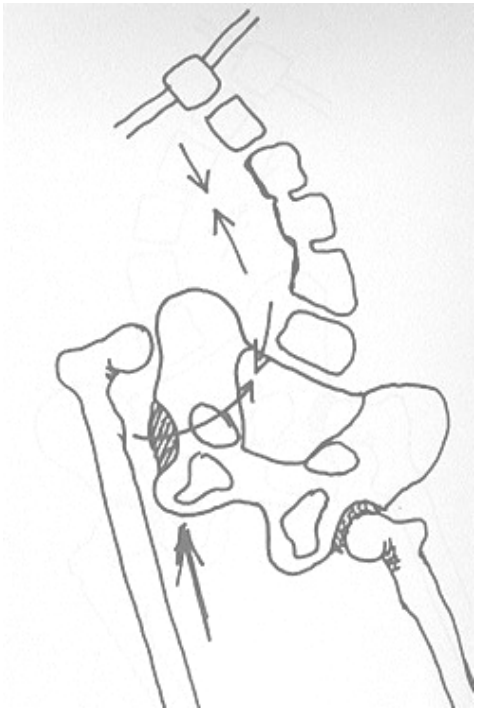
Fig 4: Frischhut classification Type 3



In the wind blown hip syndrome both scoliosis and a pelvic tilt are found in combination with unilateral dislocation of the hip (Fig.4). The dislocated hip is located next to the concavity of the scoliosis. Type three deformity is found in tetraplegic patients with a disparity in the involvement of the right and the left side of the body. As the iliopsoas muscle crosses the pelvis rising at the lumbar spine and inserting at the lesser trochanter, it plays an important role in the development of this complex deformity.

- **Type 4:** Pelvic tilt and Neurogenic/congenital scoliosis:

Fig 5: Frischhut classification Type 4



In spina bifida patients, muscle imbalance and congenital malformations are the essential causative factors of deformities of the spine. Muscle imbalance around the hip joint is a well-known condition mainly present in midlumbar lesions. The level of the lesion could be quite different on the right and the left side. Therefore, unilateral or bilateral hip dislocations can be found (Fig.5). A wide variety of combinations of spinal deformities, uni-or bilateral hip dislocations and pelvic tilt can be observed.

4) Physiopathology of the pelvic tilt

In a review of the literature, we found that pelvic tilt is analyzed from different points of view. In some papers, the pelvic tilt is assessed in association with the natural history of neuromuscular disorders and their functional impact. Other papers analyze the pelvic tilt as related to spinal deformities, or in association with hip deformities.

Fulford and Brown (19) conclude that muscular imbalance and alterations of muscular tone as well as gravity and positioning are of importance in the development of deformities such as scoliosis, pelvic tilt and hip deformities.

Pritchett (46) reports patients with severe cerebral palsy. He assesses pain, the ability to sit, pelvic tilt, spinal deformities, nursing problems and fractures. Fifty percent of the patients have undergone hip surgery while the other half has not been subject to any hip interventions. Pritchett found that the ability to sit is not related to hip deformities. The degree of scoliotic deformities and the pelvic tilt was found to be dependent on the severity of the neurological involvement and not on the hip deformity. A causal relationship between a pelvic tilt and a scoliosis has been assumed. Moreover, it is thought that there is no causal relationship between a pelvic tilt and a hip deformity.

Letts et al, (30) have observed the natural development of scoliosis, pelvic tilt and hip dislocation in 22 children with cerebral-palsied. They found dislocation of the hip was followed by a pelvic tilt and the subsequent development of a scoliosis in 73% of these children and suspect that the unilateral dominance of the adductor muscle and the iliopsoas muscles are the main causative factors. In 27% of these children a scoliotic deformity was found initially associated with unilateral muscular hyperactivity.

Howard and Mc Kibbin (26) have been able to demonstrate that there is a link between the severity of cerebral palsy and the incidence of hip deformities and pelvic tilt.

Lonstein and Beck (33) report on 464 children with cerebral palsy. They have not been able to identify a statistically significant correlation between pelvic tilt and hip dislocation. In their series unilateral or bilateral dislocation of the hip has been noticed as independent from the pelvic tilt. However, they have encountered a correlation between scoliosis and pelvic tilt: the pelvis is always lower on the convex side of the spinal deformity. One group of their patients presented with scoliosis, pelvic tilt and unilateral dislocation of the hip, a deformity that has been called windswept deformity by Letts and others (30). They have concluded that various combinations of unilateral and bilateral dislocations of the hips can be found in concurrence with a pelvic tilt and that a unilateral hip dislocation does not necessarily cause the pelvic tilt. They consider the underlying neurological lesion to be the main causative factor for deformities.

A similar observation has been made by Pritchett (47) in patients with severe cerebral palsy. A dislocation of the hip, unilateral or bilateral, will not be related to a pelvic tilt.

Samuelson and Oklof (53) have analyzed in patients with myelomeningocele the correlation between scoliosis, level of the neurological lesion, congenital spinal deformities, pelvic tilt, hip dislocation and neurological problems above the level of the neurological lesion. They have not been able to establish a correlation between scoliosis and hip dislocation. In their series relationship was found between pelvic tilt and scoliosis: the convexity of the spinal deformity is always found on the same side as the lower pelvic rim. They have concluded that muscular imbalance below the pelvis cannot be considered to be the causative factor of neither the pelvic tilt nor of the scoliosis.

Cooperman et al (12) report on 38 patients with cerebral palsy (average 26 years old) who they have followed for 18 years. They have found a correlation between hip dislocation, pelvic tilt and scoliosis mainly in cases of severe cerebral palsy. In comparing the severity of cerebral palsy, they have encountered that patients who were successfully operated on for unilateral hip dislocation presented a lesser degree of pelvic tilt and scoliosis at reassessment. They assume that reduction of the hip might result in neurophysiological alterations and recommend surgery in cases of unilateral dislocation in order to prevent both deterioration of the pelvic tilt and of the scoliosis.

Pfeil and Niethard (45) propose that an unilateral hip dislocation and asymmetrical reflex activity of the muscle around the hip are the primary causes for a pelvic tilt. More over, they presuppose that these factors cause a compensatory scoliosis that becomes structural in spina bifida patients. A correction of more than 50% of the pelvic tilt has been achieved by muscle release surgery around the hip.

Lindseth (31) has evaluated the results of surgical correction of the pelvic tilt in spina bifida patients. He has stressed that not only muscular imbalance but also congenital spinal deformities have to be considered as causative factors for a pelvic tilt.

a) Cerebral Palsy

Originally, the term Cerebral Palsy referred to birth anoxia or trauma that led to cerebral dysfunction and spasticity. Today, it is more of an all inclusive term for the static encephalopaties, whether they are caused at the prenatal, perinatal, or postnatal stage. It includes all the non-progressive brain disorders such as genetic demyelinitation, intrauterine cerebral infections (toxoplasmosis), perinatal anoxia, cerebral trauma or hemorrhage..

The spinal deformities in these patients can be separated into postural curves (caused by lack of sitting balance) and structural curves.

At first glance, the etiology of the structural scoliosis appears to be easy to explain on the basis of muscle imbalance on the two sides of the body and of insufficient dynamic stabilization against gravity.

The overall incidence of spinal deformities in cerebral palsy is of 20 to 25%. In fact, there is a low incidence of scoliosis in spastic diplegia (5%) and a high incidence (65%) in spastic quadriplegia.

Progression of scoliosis in this group of patients differs from that of idiopathic scoliosis where progression occurs mainly during the rapid growth spurt of puberty.

In cerebral palsy, the onset of puberty is variable and can occur as early as in age 8 or as late as in the early 20s. In addition, progression continues into adulthood in a high percentage of cases. This means that the patients must be kept under observation for a long time, well into adulthood. When deciding whether a specific patient should undergo spinal fusion, it is important to take into account the natural history of the scoliosis and whether his curves have progressed.

Majd and colleagues (36) have followed patients in a nursing home and have documented whether or not they have had scoliosis and whether their curves have progressed. They have found that 18% of the patients have had a significant deterioration in their curves. The larger curves have tended to progress in adulthood at a rate of 4,4° per year. Three patients have experienced decubiti and their average curves were greater than 100°, with more than 45° of pelvic obliquity.

Thometz et al (60) have similar results, but the rate of progression of curves greater than 50° at skeletal maturity has averaged only 1.4° per year. They have also registered that thoracolumbar and lumbar curves tend to progress more than thoracic curves.

b) Duchenne Muscular Dystrophy (DMD)

Scoliosis in the Duchenne group shows a variation in the progression of the curve and the subsequent prediction of spinal deformity. If the Duchenne patient presents a significant lordosis or is able to walk, scoliosis and kyphosis rarely develop.

The incidence of progressive scoliosis is about 95% in non walking patients with Duchenne muscular dystrophy. The natural history of these curves is a progression to more than 100°. It has been noted that when the patients curve exceeds 35°, the vital capacity is usually less than 40% of the predicted normal value.

Hsu et al (27) have found that the curve progressed from 0,3 to 4,5 degrees per month in patients between the ages of 13 and 22 years of age.

Three types of DMD scoliosis have been described by Oda (41):

Type 1 (progressive C lumbar curve)

Type 2 (double curve with variable progression)

Type 3 (Stable curve).

The hyper lordotic spine tends to be more stable, in contrast to a kyphotic spine. The pattern of the curve, the Cobb angle and the vital capacity at the age of ten years are parameters which have been used recently by Yamashita et al to predict the progression of the curve (64). A plateau of vital capacity of less than 1900ml has been associated with rapid progression of spinal deformity.

Pelvic obliquity is caused in part by the general imbalance of the trunk with the pelvis becoming part of the curvature. Gravity accelerates the progression of the spinal deformity and a flexion contracture of the hip leads to subluxation exacerbating the pelvic tilt (50).

Forst et al (18) have concluded that the pelvic tilt and muscular weakness are the main factor leading to sitting problems and difficulties in trunk and head control. They have recommended early surgical correction and stabilization. Moreover, they stress the fact that sitting comfort is improved with surgical spinal stabilization and cosmetic appearance is not be underestimated for improving the psychological condition of these patients.

c) Poliomyelitis:

Poliomyelitis is primarily a viral intestinal infection. However, once the central nervous system is invaded, the motoneuron cells of the anterior horns of the spinal cord and brain stem are destroyed or temporarily lose the ability to function. The earliest sign is chromatolysis of the Nissl substance in the cytoplasm. Then it is followed by inflammatory infiltration of polymorphonuclear and mononuclear cells in the perivascular regions and, later, diffuse scattering in the grey matter. The loss of function in the motoneuron may be either reversible or not. This is reflected clinically by the recovery of some muscle power in certain patients. Bodian has postulated that nerve cells may undergo slow recovery, leading to morphologic recovery within a month. In irreversible cases, chromatolysis progresses, the nucleus shrinks, and the necrotic cell is removed by neuronophagia.

Kuo et al (28) have stated that in long-term follow-up of 118 adult cases of Poliomyelitis in southern Taiwan, the most common deformity has been scoliosis (67,8%), followed by limb inequality (54,1%) and foot deformity (51,7%). Roughly, at least two thirds of poliomyelitis patients will develop evidence a scoliotic deformity**.**

The type and severity of the scoliotic curvature that develops depends on the age of the patient, the extent and residual power of the involved trunk muscles and pelvic obliquity. Thus, the curve may be in the high thoracic, thoracolumbar, or lumbar area, depending on which groups of muscles are paralyzed. The curve patterns are: double major thoracic and lumbar 55%, thoracolumbar C type 23%, lumbar 16%, thoracic 5%, double thoracic 0,3%.

The natural history of scoliosis in poliomyelitis is not as clear as that of idiopathic scoliosis. Both the outcome and the eventual severity of the curve are difficult to predict. In general, scoliosis secondary to poliomyelitis is mild and non-progressive.

The degree of limb length inequality and the presence of pelvic obliquity accentuate the deformity in the lumbar region. Pavon et al (43) have noted that leg length inequality is present in about half of their patients with pelvic obliquity.

d) Spinal Muscular Atrophy (SMA)

Spinal muscular atrophy encompasses a group of heritable conditions characterized by degeneration of the anterior horn cells of the spinal cord and, at times, the neurons of the lower bulbar motor nuclei. Clinical characteristics common to all conditions of this entity are relatively symmetrical limb and trunk weakness and muscular atrophy that affect the lower

extremities and the proximal muscles more than distal muscles. Scoliosis, joints contractures, and hip dislocations are the most common and severe orthopaedic conditions associated with this disease. It could be expected that once the diagnosis of SMA is made, most of the patients that survive into adulthood, will develop scoliosis.

The pathogenesis of the scoliosis in SMA develops as a result of muscle weakness. Frequently, severe and diffuse weakness leads to a collapsing spine and progressive deformity that may involve the entire spine and pelvis. The deformity is usually progressive and is independent of the patient's remaining growth; it may progress in spite of skeletal maturity. The presence of contractures in the pelvis and lower limbs, hip subluxation or dislocation, and the lack of equilibrium can contribute to progression of the spinal deformity.

SMA is presented in two types:

1) Type 1: Acute infantile form. The disease is present at birth or its onset is usually before the age of 2 months. These patients never walk, turn over, or sit alone and often have a bulbar paralysis, extreme muscle weakness, respiratory deficiency, recurrent pneumonia and atelectasis. Without supportive medical treatment, many of these children would die with in a few days or weeks of life.
2) Type 2: Chronic childhood form. There is a severe spinal muscular atrophy. This form of spinal muscular atrophy has been considered as a progressive disease in the past. Some authors have presently challenged this concept and reported a non progressive disorder.

Munsat et al. believe that if there is no progression of the disease for two years after the onset, further progression is unlikely.

There is a considerable controversy as to the relationship between the degree of disability and the age of the patient. Van Wijngararden (62) found no correlation between these two parameters. Barois et al (3), report 100 cases of spinal muscular atrophy that have survived beyond the age of 4. At an average follow-up of 16 years, 71 patients have never walked, and 6 have died. In the ambulatory cases, loss of walking ability has occurred in 24 patients, at an average follow-up of 20 years, while 2 have died. Pearn et al (44) have shown that no child with spinal muscular atrophy has been able to run after the age of 12. Patients with more benign disease stopped walking at approximately 20 years of age. They have reported that all patients whose clinical onset of disease occurred before they achieved normal running ability, showed progressive disability. In their study, 25% of the patients show no deterioration. Although some evidence exists of a few cases with an unusual form of the disease, progressive degeneration seems to occur in most patients with the chronic childhood form of spinal muscular atrophy.

The occurrence of spinal deformity is to be expected in most patients with spinal muscular atrophy. Daher (13) et al report that in patients who were diagnosed between 1 and 14 years of age, with an average age of 6,8 years, curve progression occurred in all patients. In the

study of Riddick et al (49), scoliosis was diagnosed at an average of 6 years of age. In their study, twenty-two scoliotic curves and four kyphotic curves progressed to an average of 95°.

There is some controversy over the relationship between motor developments, the age at onset and severity of scoliosis. Aprin et al. after studying 22 patients who had had spinal fusion, found no difference in the age at onset of the scoliosis between the group of patients who could walk and the group who could not. Conversely, Schwentker and Gibson (59) have found that only seven of fifteen walkers had scoliosis, and of those, only one patient presented a curve of more than 60°. Evans et al (16) have encountered a direct relationship between functional motor developments, the age at onset and the severity of the spinal deformity. Patients with more severe muscle weakness exhibit deformity as early as 2 years of age, whereas patients with less involvement present at a mean age of 8.4 years. Only in the group of patients who developed the ability to walk or run, were there cases with no spinal deformity. However, even in this group, 7 out of the 11 patients were found to have some spinal deformity.

Summarizing these studies, it appears that the mean age at onset of scoliosis is somewhere between 5 and 9 years. Patients that preserve the capacity to walk are unlikely to develop severe scoliosis (23).

5) Treatments

A) Conservative (Brace, sitting support)

a) Cerebral Palsy

Non-operative treatment of neurological scoliosis with adaptive seating and orthoses can help in maintaining a well balanced spine, but will not alter the progression of the deformity. (66)

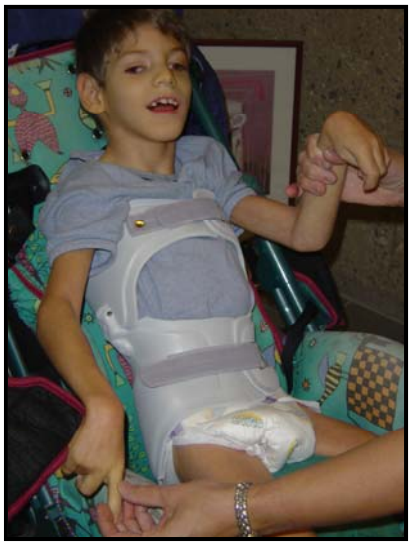
In cases in which support is necessary, because of lack of balance when sitting - two options are available:

- Thoracolumbar brace.

Bracing had no impact on the curve, shape, or rate of progression of the scoliosis in spastic quadriplegic patients who were followed by Miller and colleagues (38). Bracing can lead to skin intolerance in these thin children but Letts et al (29) have found that seating is made somewhat easier when a soft orthosis is prescribed (Fig.6). In this case the brace was used only to allow comfortable seating, not to treat the curve.

This brace is made in soft material with a polyethylene core to reinforce the brace. It helps both positioning and transfers of the patient. This orthosis is popular among patients and parents at our clinic.

Fig 6: Thoraco-lumbar brace



- Moulded sitting support.

The molded polypropylene shell is made from a positive mould of the patient. It is a total-contact orthosis, with support from the distal posterior thighs to the shoulders or occiput, where necessary. This orthosis gives excellent pelvic and thoracic control, there are hardly any skin problems (Fig.7,8). A head support may be added as required to control the tendency of the head to flex. In addition, the feet are placed on footrests to avoid too much popliteal pressure. In all sitting supports, a lap belt is mandatory to prevent the pelvis from sliding forward.

Drummond et al. describe several prerequisites to prevent the patient developing a pressure sore.

1) Not more than 30% of weight distribution should be on the ischium,
2) Combined ischial and sacrococcygeal pressures, should not exceed 55% of the patient's weight. Sacrocoxigeal pressures alone should not exceed 11%.
3) Non ambulatory patients have a greater risk of developing an ulcer.

Fig 7: Pressure sitting on a table (67). The red spot show the hyper pression in the ischium when sitting with out a correct support

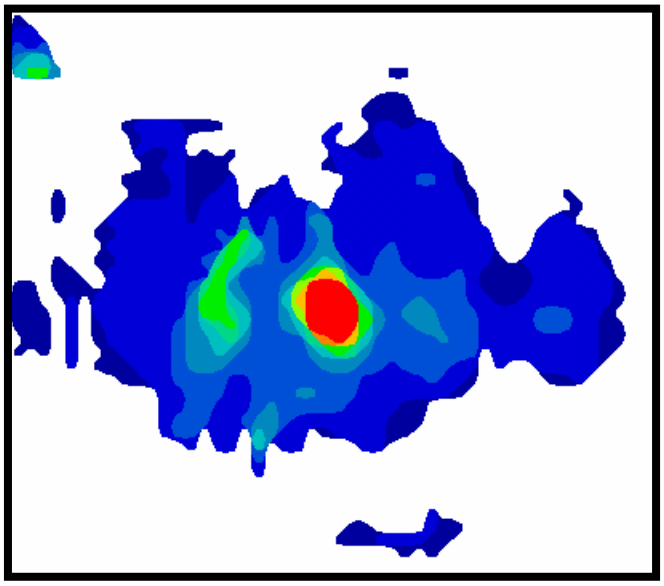
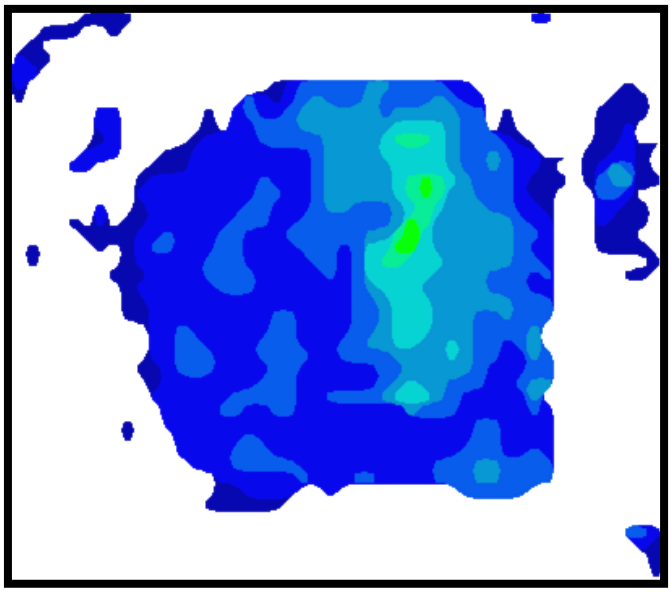


Fig 8: Pressure sitting on a moulded cushion. We can see that the hyper pressure has been well distributed.



Wheelchair adaptation can help the child with scoliosis sit more comfortably, but has little effect on worsening the curvature.

Some guidelines that let us adapt the wheelchair to the pelvic obliquity:

We consider wheelchair adaptation as a body orthosis on wheels. Wheelchairs should have a stable support, allow to lean forward, support the back, freedom of the feet, security and good pressure distribution. On this last point it is very important to adapt the support to pelvic obliquity. Besides providing the stability required, the seat must not cause uncomfortable pressure. Pressure is what creates stability; and it should be distributed in a larger surface. In contrast, if it is concentrated on a small surface of the body, it decreases stability.

Discomfort due to pressure results in muscular fatigue when the user tries to steady the instability of the trunk. Pressure distribution over a large, soft area gives more comfort, but if the area is too soft, stability decreases.

The ischial tuberosities are the bones on which we sit. Propioception from the ischial tuberosities and the surrounding tissue is important for balance. The sacrum and coccyx shouldn't have pressure on them because serious painful problems may occur.

The hip region is always problematic when pelvic obliquity has developed. There is a high risk of developing pressure sores in this area if this is not taken into account when making the seat cushion. Buttock muscle and soft tissue function as internal "seat cushions", but sitting for too long blocks the circulation of blood and the tissue becomes temporarily numb, longer lasting, static pressure can cause severe ischaemic pain.

b) Duchenne Muscular Dystrophy:

The management of the spinal deformity in Duchenne's muscular dystrophy (DMD) has evolved considerably since the early 1970s. Spinal ortheses were said not to control the curve

in 94% of patients. Curve progression was noted even in the sample of 32 boys with curves of more than 30 degrees who wore braces (11). If scoliosis does develop while the child is still able to walk, bracing will be ineffective and will probably make him non-ambulatory.

c) Poliomyelitis:

A thoraco lumbosacral orthosis (TLSO) for the trunk may be cumbersome and may interfere with the patient's already weak respiratory muscles in the early stages of Poliomyelitis. The long-term use of a TLSO is inconvenient and detrimental in growing children, as it further weakens the trunk muscles.

The extent of muscle involvement, the degree of motor power, and any deformity, joint contracture, or limb length inequality must be measured and recorded. The presence of pelvic obliquity and the ambulatory status of the patient (independent walker, crutch-assisted walker, or wheelchair bound patient) are significant for determining the optimal methods and time of treatment of the joint and spine problems.

To date, serial casting, brace application, and surface electrical muscular stimulation have been ineffective in controlling or correcting spinal deformity in this group of patients. Physiotherapy or muscle training also fails to reduce or even stabilize curve magnitude. Braces may be used in young children with collapsing spines waiting while for the spine to be mature enough to perform spinal fusion.

d) Spinal muscular atrophy

Orthotic treatment may be prescribed with the objective of reducing the rate of progression of the spinal deformity or of helping to maintain the sitting posture.

Evans et al have reported the results of orthotic treatment in adult patients with severe deformity to avoid painful impingement of the lower ribs on the iliac crest. The authors' opinion is that orthotic treatment will not avoid the development or ultimate progression of the deformity but may support the spine for a time. Schwentker and Gibson (59) found that the use of a brace did not prevent the subsequent development of scoliosis. Patients who used a brace before the diagnosis of scoliosis, they developed deformity all the same. Shapiro and Bresnan (54) have written that a brace can be prescribed when a curve reaches 15 to 20 degrees to minimize the rate of progression of the scoliosis. Riddick et al (49) reported similar results on 20 patients who received conservative treatment. Although half of their patients have only been followed for two years or less, their data suggests that the use of a brace was effective in slowing curve progression.

Bracing is recommended for use in children younger than 9 years old, when the deformity reaches 20°, to decrease the rate of progression. This treatment allows spinal fusion to be delayed until the child reaches 10 years of age or more or the curve attains 50°. Granata et al (23) have not found braces useful in preventing scoliosis, but the orthosis improved the functional performance of all children.

In a young child with a severe acute form of SMA and poor control of the neck, a sitting support orthosis is utilized. A moulded plastic posterior shell with control from the head of the pelvis is useful and can have a separate removable anterior shell or straps. The purpose is to support the spine and thus provide sitting ability.

For a less affected child, a lightweight three point pressure brace can be used. For most of the patients the thoraco-lumbosacral orthesis is a good option.

B) Surgery

a) Cerebral Palsy

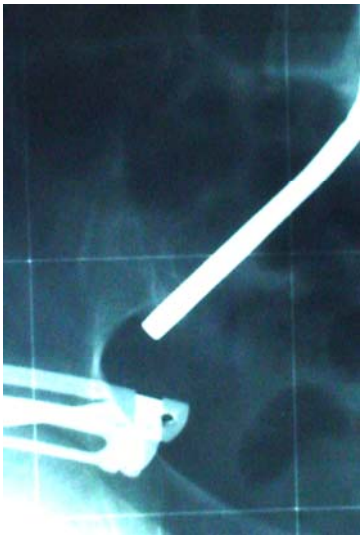
With such discouraging results from non-operative treatment, it would seem to be logical that patients with scoliosis due to cerebral palsy would be offered surgical correction. Yet, this is not the case. Spinal fusion surgery carries sufficient risks that it is not considered to be in the best interest of every child with neuromuscular scoliosis.

In this group of patients, the two main indications for surgery are documented curve progression and deterioration in function. Otherwise, a formal contraindication for surgery is the absence of vision and no contact at all with the world that surrounds him.

In the preoperative assessment, evaluation of the child’s health is important. A multidisciplinary team approach to the patient should be used. The two areas of greatest

importance are the respiratory assessment and an evaluation of the nutritional status. When the later is not optimal, a feeding gastrostomy is often placed and surgery postponed until nutrition is improved.

Fig 9: Galvestone rod perforating the iliac crest



The surgical management of the spinal deformity in patients with neurological disorder has changed in the past few years. The use of the Harrington Rod distraction technique resulted in many complications (Fig.9) (Pseudoarthrosis, rod fracture, failures of hardware fixation, brace complication with skin ulcers) (9, 34).

Luque segmental instrumentation with sublaminar wires was introduced in the late 1970s. The L shaped Luque double rods pose the problem of vertical shift and rotation, and a coupled single-unit rod has therefore been introduced and widely used in the 1980s to counter these forces (37).

In 1982 a technique using intrailiac rods (Galveston) for the correction of pelvic obliquity (1) was developed and in 1984 the Cotrel Dubousset and Texas Scottish Rite Hospital systems which use laminar hooks and pedicle screws, were introduced.

The Isola system uses a combination of fixation by pedicle screws into the lumbar spine, hooks and wires. This has been reported to give good correction of scoliosis and pelvic obliquity when used with Galveston pelvic extension rods (65, 24).

The trend in the 1980s has been to extend the fixation to the pelvis or sacrum. The main aim of correcting the pelvic obliquity has been to facilitate the erect sitting position in a wheelchair, without pressure sores. Lonstein (32), in keeping with many other authors, suggests that in Type 2 curves (Lonstein classification) that whether or not the pelvis obliquity is included in the lumbar curve, the pelvis should be included in the fusion area. Mubarak (39) accepts up to 10° before including the sacrum in the fusion area and recently Gaine et al (20) accepted up to 30° of pelvic obliquity.

The length of the surgical procedure as well as the increased blood loss has persuaded many to limit the surgery to the lumbar spine and not to the sacrum.

Fusion only to L3/4 risks retrolisthesis and translation of the L5 vertebra with persistent pain (24).

At the Geneva University Hospital fusion of the lumbar curve up to L5 is undertaken when the sacrum is not included in the lumbar curve.

b) Duchenne Muscular Dystrophy

In the DMD group, the timing of fusion is dictated by the degree of curvature and pulmonary function. Since 95% of curves progress once the patients are wheelchair bound, the trend over

the last decade has been to fuse early (57). The benefits of surgery in this group include improved sitting in a wheelchair, more comfort when lying and improved respiratory excursion (15).

Prolonged life expectancy is a potential benefit. When comparing the survival data, a significantly higher mortality is observed in those patients who have refused surgery, indicating that spinal stabilization can improve survival for several years following operation.

With regard to the surgical procedure, as far as the optimal level of fusion is concerned, for patients in whom fusion was performed no lower than L4, the outcome was less successful in terms of correction of the curve and maintenance of the correction.

Most authors in North America encourage pelvic fixation. However, fusion to the sacrum or L5 has produced a similar long term correction of the pelvic obliquity.

Forst (18) recommends early instrumentation of the sacrum in order to prevent pelvic obliquity.

A combined anterior and posterior fusion in one or two stages is the most effective way of correcting pelvic tilt in other neuromuscular disorders but this type of combined approach poses a high risk of blood loss and hypotension in DMD.

Fixation of an intrailiac pelvic rod achieves the best immediate correction of the pelvic obliquity. The disadvantages of instrumenting the pelvis include a longer operating time, greater blood loss, more difficult surgery (technically), and risk of non-union because of the

severe osteoporosis and pelvic pain. For these reasons, Mubarak (39) suggests to arthrodese the spine only to L5 level, unless the pelvic obliquity is greater than 10° or the scoliosis curve greater than 40°.

Although pelvic fixation has given the greatest immediate correction of the Cobb angle and pelvic obliquity correction, fusion to L5 has a similar long term effect and maintenance of the obliquity. Also Gaine (20) states that fusion to levels L5, S1 or the pelvis have given comparable results in the scoliosis correction with an initial curve in the order of 50° being maintained at 35° at four years follow-up.

With the use of a respiratory support system and steroid supplements, the life expectancy of patients with DMD is likely to increase into the late twenties and early thirties .That is why long-term maintenance of correction of spinal and pelvic deformity has become more crucial. Rates of complications vary from 32% to 61% (58). Major complications including cardiopulmonary failures, infection and problems with hardware have occurred in 27% of 30 patients reported by Ramirez (48).

In summary, the literature for the DMD group shows that fusion to L3 or L4 gives inadequate correction. It should be recognized that some pelvic obliquity is acceptable, possibly up to 30°, with a balanced upper trunk and appropriately adapted seating (20).

c) Poliomyelitis

Curve progression is the main reason for patients to seek treatment.

In patients with lumbar scoliosis, pelvic tilting can be relentless, causing severe trunk deviation and unilateral ischial sitting. The goal of treating spinal deformity is to obtain a vertical torso centered over a leveled pelvis. This allows for stable sitting on level buttocks and helps prevent ischial pressures callosities as well as paralytic dislocation of the hip on the high side of the pelvis. The patient does not have to use the arms for support and is able to use both hands freely to perform other activities.

When the spinal curvature is severe or is collapsing, the lower costal margin sometimes impinges against the upper margin of the iliac crest, causing pain. Respiratory capacity and ambulation are also hampered. In this situation, a straight and stable spine can relieve symptoms and facilitate standing and walking. Back pain may be a major problem in adult polio patients with scoliosis.

Surgery is considered only when the Cobb angle is larger than 50°, when the spine is collapsing or severely unbalanced.

Posterior instrumentation alone is indicated for thoracolumbar or lumbar curves of less than 70°. If the curve is larger, whether flexible or rigid, or pelvic obliquity is severe, a two stage surgical procedure is recommended (anterior liberation and posterior instrumentation), as this results in better correction of the deformity and less pseudoarthrosis. Halo traction can be

used in severe scoliosis pre-operatively or between the anterior and posterior approaches. It can be utilized for up to two weeks, with the maximum correction achieved at that time.

If pelvic obliquity is to be rectified, the fixation should extend to the pelvic region as has been traditionally stated.

Caution is also recommended, within this group, when fusing to S1 because walking may be affected.

d) Spinal Muscular Atrophy

The usual indication for an anterior procedure in a neuromuscular scoliosis is a severe rigid curve with a pelvis that cannot be leveled or a torso that cannot be placed in balance over the pelvis by external maneuvers. In children with spinal muscular atrophy, there are some other factors to be considered. These children are more dependent on the diaphragm and abdominal muscles for breathing than some patients with other neuromuscular disease because of the weakness of the muscle in the neck and thorax. Anterior approach and fusion, especially when done through a thoraco abdominal approach, decreases the respiratory and coughing capacities of such patients - with a subsequent increase in pulmonary complications.

Before indicating operative treatments in SMA, the following should be taken into account:

a) In the chronic form of the spinal muscular atrophy, neurological progression of the disease is slow.

b) Spinal deformity almost always develops and will progress, regardless of any non-operative treatment.

c) Severe scoliosis leads to a poor sitting posture, which diminishes the patient's functional and pulmonary capacities.

d) Deterioration of the pulmonary function progresses with the age of the patients.

The goal of the surgical treatment is to have an arthrodesed spine over a leveled pelvis. With these considerations in mind, a rational approach is to perform posterior spinal fusion before the deformity becomes severe and the pulmonary function becomes restricted. At this primary stage, the surgical procedure is less extensive, allows better correction, and lessens post operative complications.

In a severe deformity, the rate of complications increases with the severity of the curve and the goal of achieving a stabilized spine centered over the pelvis must be weighted against the risks involved.

Most authors recommend posterior spinal fusion at moderate deformity after the age of 10 years.

As was mentioned before, orthotic treatment will not avoid the development or ultimate progression of the deformity but may support the spine for some time**.** Riddick et al (49) have reported 20 patients who have used a brace; this has been effective in slowing curve progression. Curve reduction and compliance with the brace are two factors that influence the progression of the scoliosis.

Shapiro et al recommend surgical treatment when the curve is between 40 and 60 degrees. They point out that when the curve exceeds 100°; anterior fusion is required, adding considerable risk. Others authors assume that the fusion should be performed when the curve is situated between 30° and 60°.

Patients scheduled for surgery benefit from comprehensive evaluation by a pulmonologist, a neurologist, an anaesthesiologist, and a physical therapist. Assessment of the pulmonary function a few weeks before the operation provides a useful and relatively simple baseline of the pre-operative condition. In a study done at the Alfred Dupont Institute (42) involving patients who underwent posterior spinal fusion for neuromuscular scoliosis, it has been observed that patients with a vital capacity of 50% or less of the predicted value were at risk for the development of pulmonary edema or atelectasis. Patients with a vital capacity of more than 60% did not have pulmonary complications following the surgical procedure. Patients with a vital capacity of less than 25% were at severe risk and required extensive therapy.

In spinal muscular atrophy, as in others forms of neuromuscular scoliosis, the function of instrumentation is to produce safe correction of the deformity, allow fixation from the upper thoracic spine to the pelvis, and provide rigid fixation that eliminates the need for post-operative casting or traction. Segmental fixation with Luque rod or the unit rod is widespread in the treatment of neuromuscular scoliosis. These systems provide rigid fixation and usually do not require post-operative external immobilization. Correction of pelvic obliquity and

decompensation of the torso are more effective with the use of these systems and the intrailiac fixation described by Allen and Ferguson. This is important, because pelvic obliquity places excessive weight on a single ischium, which limits the ability to sit for long periods.

6) Materials and methods:

We reviewed the charts of patients with neurological disorders that had spinal fusion up to L5 or S1 from 1985 to 2001, with a follow-up of more than two years and with full charts and X-rays available.

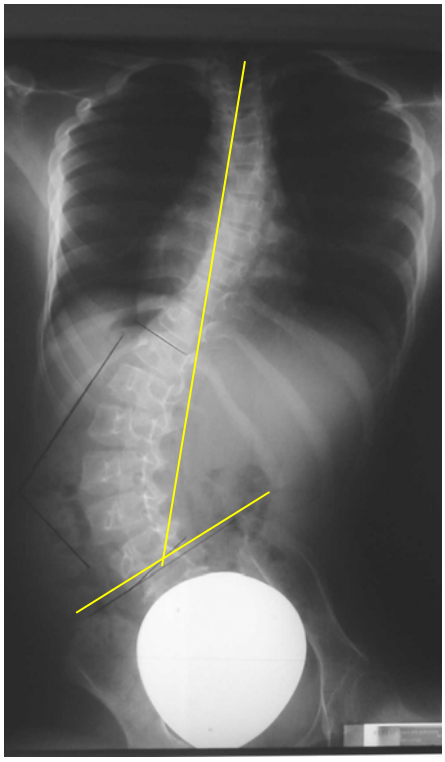
The hospital and office charts as well as radiographs of all patients have been assessed. We have recorded the curves in the coronal and sagittal plane (pre-operatively, post-operatively, and at the time of last follow-up), fusion status and complications.

All curves in the sagittal and frontal plane were measured using the Cobb method. Scoliosis, kyphosis, lordosis and pelvic obliquity have been measured on an upright radiograph before the operation, after the operation, and at the time of latest follow-up.

X-rays were taken standing or sitting according to the functional status. In patients with poor balance, the upright radiograph was taken with minimal support in order to show the maximum curvature and pelvic obliquity. Any radiolucency surrounding the pelvic rods was measured on X-ray.

Pelvic obliquity was measured on antero-posterior radiographs as described by Maloney (37), using a line drawn across the superior aspect of both iliac crests and a second line drawn from the center of T1 to the center of S1 (Fig.10). The pre-operative X-ray, immediate post-operative and last follow-up radiographs were chosen, all of them in sitting or standing position.

Fig 10: Measurement of pelvic obliquity with the angle between T1-S1 line and the biiliac crest line (33)



Only patients with fusion to the level L5 or S1 have been included. If the fusion level stops above L5 the patient was excluded from this study.

The indications for the surgical procedure included a deformity that affected the ability to sit continuously, to breathe, to eat, to remain comfortable, threat to the integrity of the skin surface involved in sitting, or risk of the progression of the disease to the point that these functions would be affected. Curve progression documented on serial radiographs and the magnitudes of the curve were measured.

The indication to perform a posterior procedure alone versus an anterior-posterior procedure was based on skeletal age, diagnosis, maturity, pelvic obliquity, and flexibility of the curve noted on preoperative bending films (68). The major indications for two-stage operations are based on bending X-ray: >50°, a fixed curvature in the sagittal plane (kyphosis or lordosis),

and lumbar scoliosis with fixed pelvic obliquity.

Fusing the spine to the sacrum is indicated when the pelvic obliquity is included in the lumbar curve (Type 2B from Lonstein classification).

Type of fusion and type of instrumentation used for the spine fusion were also recorded.

In the charts, we reviewed several factors: pain at lumbosacral area that needs medications, chair adaptation (more than 2 years between adaptations has been taken as a bad result), walking ability and surgical complications.

To assess the patient's functional ability, a modified Rancho Los Amigos classification (5) has been established:

- Group 1: patients that are independent walkers or community ambulators
- Group 2: household or physical therapy ambulators
- Group 3 : independent sitters
- Group 4 :dependent sitters
- Group 5: non-sitters or bedridden patients

All patients were examined, and the clinical records, including radiographs, were reviewed.

Patients with myelomeningocele and congenital scoliosis were not included in our study because they are not considered comparable with the other patients with neuromuscular scoliosis. In Myelomeningocele patients with sensation deficit can't accept a pelvic obliquity because pressure sores can easily become ulcers. We agree with Drummond who suggests that pelvic obliquity can not be accepted in the group of patients without sensation due to a traumatic or a congenital aetiology. Congenital scoliosis -even accompanied with neurological symptoms- is a different pathology with a different management.

a) Statistical analysis:

After testing the normality of the data with the Jarque-Bera test and the equality of the variance, we analyzed if there were differences between the two surgical conditions (spinal fusion to L5 and to S1) with the t-tests. The Matlab Statical ToolBox 7.2 was used .The level of significance was set at $P< 0.05$.

6) Results

Our research -ranging from 1985 to 2001- records 31 patients with full charts available for the study.

The group comprises 15 males and 16 females with the diagnosis describe below (table 1).

Average age at surgery was 12,7 years (7 minimum and 15,8 maximum) and the average duration of follow-up was 6,58 years (2.1 minimum and 16.6 maximum).

Of the 31 cases considered, 19 have undergone spinal fusion to L5 and 12 to S1.

Taking into account the age at surgery: patients with fusion to L5 were 12.7 years old on average (+/- 2,42) and to S1 were 12,8 years old on average (+/- 1,92). There was no significant difference between these two groups. (P=0,9561)

Table 1: Diagnosis

Diagnosis	Fusion to L5	Fusion to S1
Duchenne	5	1
Polio	5	5
Spinal muscular atrophy	2	4
Quadriplegic	3	1
Friedreich Ataxia		1
Rett Syndrome	1	
Myopathy	1	
Larsen Syndrome*	1	
Syndrome with out diagnosis	1	

* Larsen Syndrome: Larsen et al. (1950) described a syndrome of multiple congenital dislocations and characteristic facies (prominent forehead, depressed nasal bridge, wide-spaced eyes).

Table 2: Surgical duration, intubation time, hospital stay and ICU stay

	Surgical duration	Intubation time	Hospital stay	ICU stay
Fusion to S1	5.78 (4.5-8) hs	24.27 hs (7-38)	17.5 (11-22) days	2.9 (1-8) days
Fusion to L5	5.35 (3-7.5) hs	18.66 hs (120-6)	17.3 (8-65) days	2.55 (1-8) days
Duchene	4.7 hs	21.2 hs	13.8 days	3.4 days
Polio	5.9 hs	12 hs	21.5 days	2.1 days
Quadriplegic	5.3 hs	12.6 hs	12.6 days	1.6 days
Spinal Muscular Atrophy	5.9 hs	26.5 hs	18 days	3.1 days

The time of surgery was shorter when S1 was not included in the fusion area (Table 2). When comparing the different diagnosis, the Duchene group has the shortest surgical duration which can be explained by the fact that most of these patients had smaller preoperative curves.

When comparing intubation time, again it is shorter when S1 is not included in the fusion area. The Duchene and the SMA groups have the longest intubation times and ICU stays because of their cardiac and pulmonary implications.

Polio patients have the longest hospitalizations. This longer stay is related to the fact that most of these patients come from Africa and thus, need to stay for longer to benefit from the rehabilitation program before going back home.

Braces were used in most of the patients with SMA to postpone spinal surgery. In the Polio group, 3 out of 10 patients used the brace in an attempt to stabilize the spine and the pelvis and thus to help with mobility.

Type of fusion technique to L5: a) CD with Screws: 10

b) Luque: 8

c) Harri-Luque: 1

S1: a) CD with screws to S1: 11

b) Luque Galvestone: 1

In two cases an anterior approach was undertaken: one was fused to S1 and the other to L5.

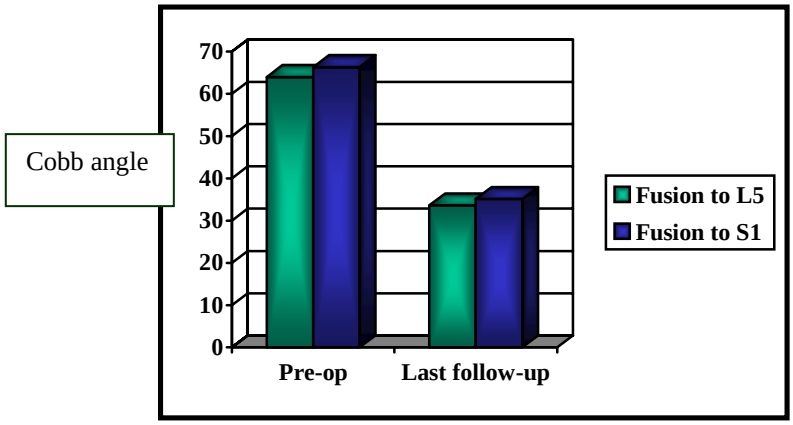
Taking into account the fusion levels, we fused 14,47 (+/-1,83) levels (per patient) to L5 and 13.8 (+/-2.48) levels to S1. There was no significant difference between these two groups. (P=0,4159)

The mean pre-operative Cobb angle was 63,94° (+/-32,87) for the patients who were fused to L5 and 66,33° (+/-27,17) for the patients who were fused to S1. There was no significant difference between the pre operative Cobb angle between these two groups.(P=0.835)

At last follow-up, the L5 group had a scoliosis that measured 30.26°, with a 52.6% correction while the S1 group showed almost the same values with a scoliosis measuring 31,08° with 53.12% of correction, when pre-operative and last follow-up X-rays were compared. Both groups showed a significant improvement of the scoliosis with a P= 0.0043 for L5 fusion and

P=0.0012 for S1 fusion (Fig.11). When we compared the Cobb angle at last follow up between both groups, the difference was not significant. (P=0.9)

Fig 11: Scoliosis Correction measured with the Cobb angle. Comparing pre-op and last follow-up



The average pre-operative pelvic obliquity for the group with fusion to L5 was 21,5° (+-18.24) and it was reduced to 18.95° (+/- 13.28) at last follow-up(Fig.12,13). (11.9%)

In the group with fusion to S1 the average pre-operative pelvic obliquity was 29,6° (+-20.49) and it was reduced to 13.5° (+/- 10.63) at last follow-up (Fig.14). (54.4%)

When we compared the pre-operative pelvic obliquity between both groups, there was not a significant difference (P=0.2426), but when we compared the amount of correction of the pre-operative pelvic obliquity and at last follow up:

1) The group that stopped at L5, didn't show a significant difference (P=0.672).

2) The group that stopped at S1 showed a significant difference (P=0.016)

Fig 12: Pelvic obliquity, comparing pre-operative and last follow-up values. Measurement of pelvic obliquity with the angle between T1-S1 line and the biiliac crest line (33)

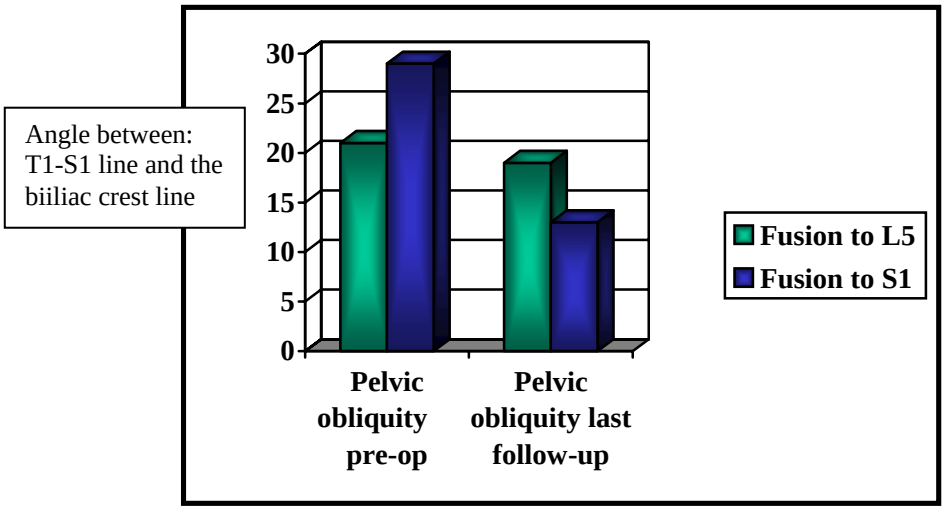


Fig 13: Individual values for pelvic obliquity correction comparing pre-op and last follow-up in the patients with spinal fusion to L5. Patient N°15 had distal loosening of fixation. Average correction: 2,5° (11,9%). Not significant difference (P=0.627). Measurement of pelvic obliquity with the angle between T1-S1 line and the biiliac crest line (33)

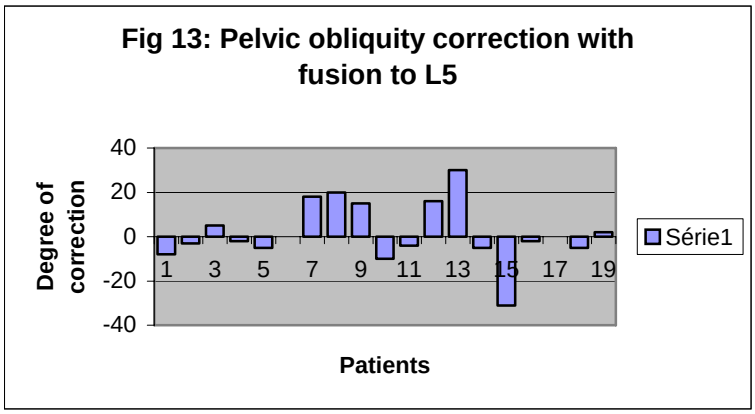
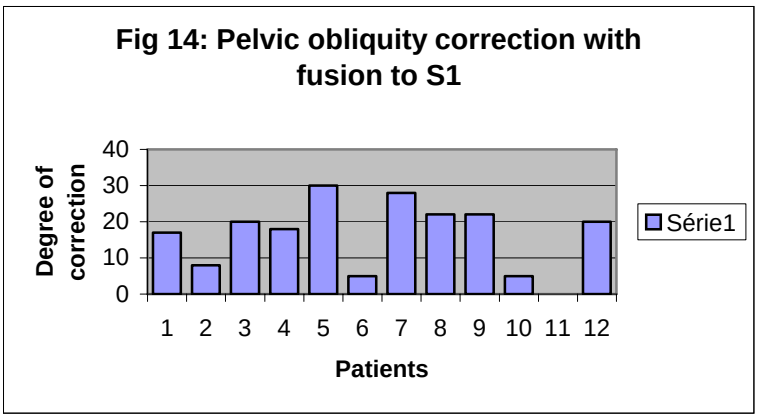


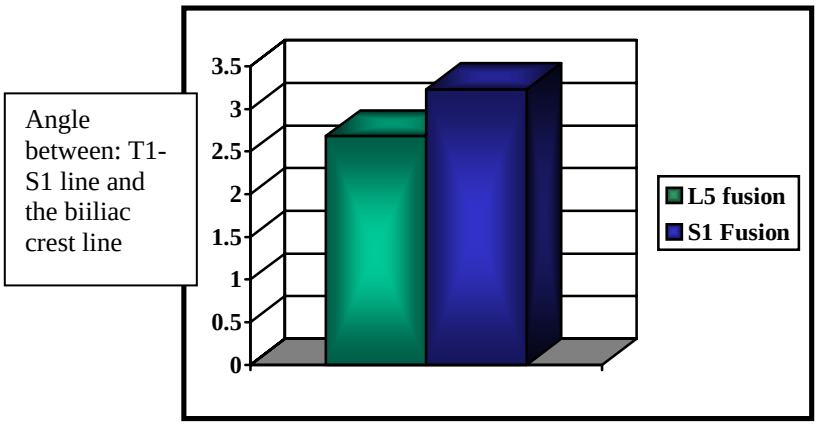
Fig 14: Individual values for pelvic obliquity correction comparing pre-op and last follow-up in the patients with spinal fusion to S1. Average correction: 16,13° (54,4%). Significant

difference (P=0.0168). Measurement of pelvic obliquity with the angle between T1-S1 line and the biiliac crest line (33)



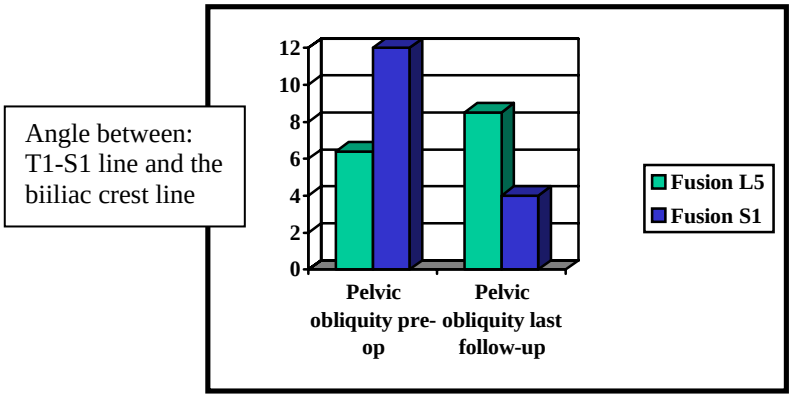
Comparing pelvic obliquity at initial x-Ray post surgery and last follow-up, the average of loosening correction for the group with fusion to L5 was 2,68° and for the group with fusion to S1 was 3,23° (Fig.15).

Fig 15: Pelvic obliquity loss of correction between the initial x-Ray post surgery and last follow-up. Measurement of pelvic obliquity with the angle between T1-S1 line and the biiliac crest line (33)



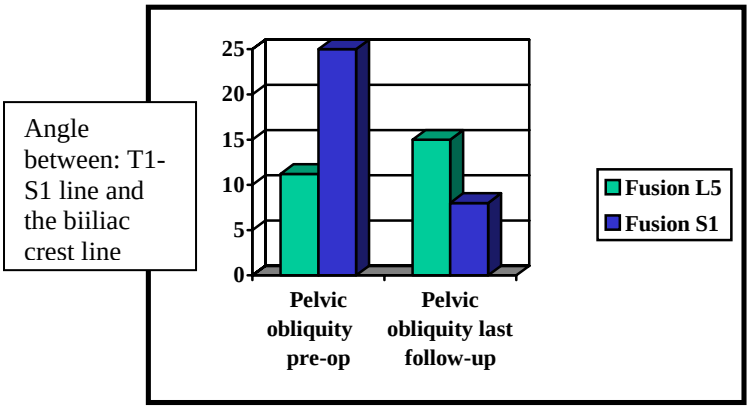
In the 6 cases with DMD (Fig.16), one has been fused to S1 with 12° of pelvic obliquity before surgery and 4° at final follow-up and the 5 others were fused to L5 with a pelvic obliquity of 6,4° (+-2,6) before surgery, 5° (+-6,7) at immediate post-op and 8,5° (+-2,3) at last follow-up. None of these cases required a second operation nor had problems with their pelvic obliquity at last follow up.

Fig 16: Duchene Muscular Dystrophy: 5 patients fused to L5 and 1 patient to S1
Measurement of pelvic obliquity with the angle between T1-S1 line and the biiliac crest line (33)



In the 4 cases with quadriplegia 3 were fused to L5, with 11,2°of pelvic obliquity pre-operatively and 15° at last follow-up (Fig.17). One case was fused to S1 and the pre-op pelvic obliquity was 25° and at last follow-up it attained 8°. In this group, none of the patients under study had any problems related to pelvic obliquity.

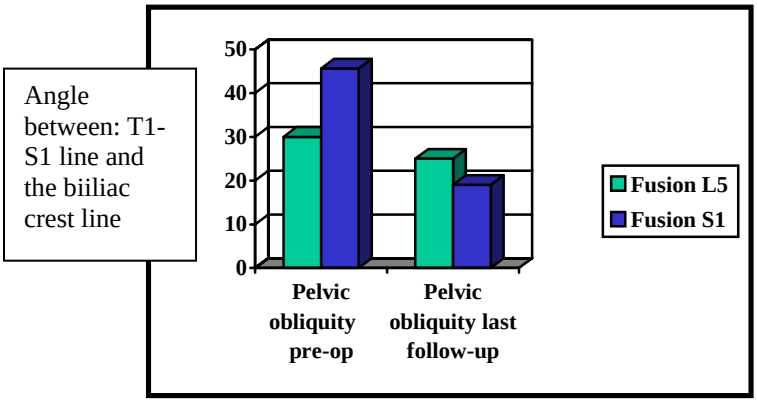
Fig 17: Quadriplegic: 3 patients fused to L5 and 1 to S1. Measurement of pelvic obliquity with the angle between T1-S1 line and the biiliac crest line (33)



In the results spinal muscular atrophy group, 2, patients were fused to L5 with 30° pre-operative pelvic obliquity and 25° pelvic obliquity at last follow-up (Fig.18). In this group, we had no correction of the pelvis obliquity after fusion of the spine to L5.

It is worth noting that one of the two patients fused to L5 has suffered subsequent lumbosacral pain. On the other hand, the 4 patients that were fused up to S1 experienced a pre-operative pelvic obliquity of 45,6° which the last follow-up was 19°.

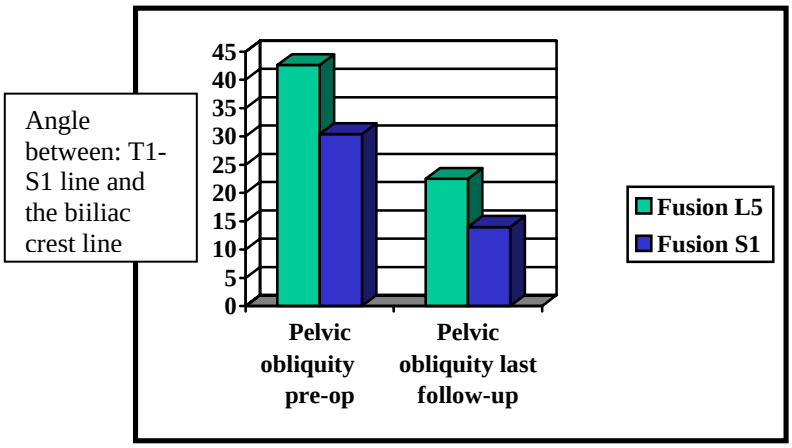
Fig 18: Spinal muscular atrophy; 2 patients fused to L5 and 4 patients fused to S1. Measurement of pelvic obliquity with the angle between T1-S1 line and the biiliac crest line (33)



The poliomyelitis group included 10 patients, 5 of whom were fused to L5 with a pelvic obliquity of 42,6° pre-operatively that was reduced to 22,5° at last follow-up (Fig.19). The other five patients were fused to S1, starting with a pelvic obliquity of 30,4° that was reduced to 14° at last follow-up. As we can see, the percentage of correction is similar in both groups. It is important to point out, that only in the polio group did we achieve more than 50% correction of pelvic obliquity when fusing to L5.

In our study, 3 patients with poliomyelitis were ambulatory patients before fusion to S1 and they remained so after surgery.

Fig 19: Poliomyelitis; 5 patients fused to L5 and 5 patients fused to S1. Measurement of pelvic obliquity with the angle between T1-S1 line and the biiliac crest line (33)



We found in relation to the neuromuscular scoliosis, 18 dislocated hips, 5 were bilateral dislocations and 8 unilateral ones. The unilateral dislocations were situated on the side of the concavity of the lumbar curve and the higher side of the pelvis. We found a relationship between the lumbar curve, the pelvic obliquity and the dislocated hip. Letts (30) described similar results in his paper.

Lonstein and Beck (33) analyzed the prevalence of pelvic obliquity and scoliosis and unilateral hip dislocation. They found that the proportion of dislocated hips is not related to the pelvic obliquity and that the direction of pelvic obliquity does not correlate with the affected hip. Scoliosis does correlate with pelvic obliquity, but the direction of the curve does not correlate with the side of the hip dislocation.

Recently, Black and Griffin (68) publish a report consistency with our findings: hip subluxation is related to the forces relating to pelvic obliquity. In 21 patients with pelvic obliquity, unilateral subluxation occurred on the high side in 17 patients and on the low side

in 4 patients. We conclude that there is a strong tendency for unilateral hip subluxation to occur in conjunction with pelvic obliquity, with most dislocations on the high side of the pelvis.

Regarding the level of independence of the different groups the modified Rancho Los Amigos classification (5) has been established:

L5 group: 12 sitters; 6 home ambulators and 1 community ambulator.

S1 group: 9 sitters, 3 home ambulators.

There have been no changes comparing the level of independence at pre op and last follow up.

From the mobility point of view, considering the 10 patients that had some degree of ambulation, this was maintained after surgery (7 with fusion at L5 and 3 with fusion at S1).

When considering chair adaptation, of the whole group of 31 patients, only one needs frequent chairs adaptation (less than 2 years) related to the pelvic obliquity and he was fused to L5:

- A myopathic patient with 55° of pelvic obliquity. During the surgical procedure, there was an attempt to fuse the spine to the sacrum, but because of osteopenic bone, good fixation could not be obtained. The pelvis obliquity did not stabilize after surgery, it continued to progress, needing regular chair adaptation to avoid pressure sores.

When considering pain at the lumbosacral area, of the whole group of 31 patients, only one had this complication that could be resolved with pain medication. She was also fused up to L5.

-A spinal muscular atrophy type 2 with lumbosacral pain, with 25° of pelvic obliquity. The fusion was taken to L5. However, it could be observed on the X-ray that part of the fusion extended to S1 with a line of pseudoarthrosis. In our opinion, the pain was related to this fact and not to the 25° of pelvic obliquity.

Complications: One loosening of fixation to L5 with re-operation to S1 and one Luque Galveston with protruding tip on the ilium. There were no infections, no deaths and no further neurological lesions in these 31 cases.

8) Discussion

Patients with neuromuscular disorders encounter a high risk of having spinal deformities (22, 32, 5). The incidence ranges from 6,5% to 67% (2, 25, 51) and these deformities are often seen together with a long, sweeping, collapsing curve, pelvic obliquity (60%) and loss of sitting balance which require, most of the time, reconstruction and fusion to the sacrum.

When we look at the scoliosis correction and at the level of fusion (to L5 or S1), the extent of reduction of the deformity is similar.

Gaine et al. (20) state that fusion to levels L5, S1 or the ileum have given comparable results in the scoliosis correction with an initial curve of around 50° being maintained at 35° at four years follow-up. In line with this result, our study shows that the scoliosis correction achieved -when fusion is taken up to L5 or to S1- is also above 50% in both cases.

The incidence of complications described in the literature is between 10 and 15% when the lumbo sacral junction is fused (1, 21). These writers mention loosening of fixation, rod mal position, pseudoarthrosis and broken rod (we mention only the complications related to L5-S1 fusion).

The description of the Galvestone technique done by Allen and Ferguson shows results with 10% of negative outcomes. Their study includes two cases with pseudo arthrosis of the lumbosacral junction, one rod fracture in the lumbo sacral junction and one rod mal position from a group of 40 patients.

Yaszici et al. mention only two protruding tips in the ilium out of 47 patients. Maloney, out of 10 patients fused with unit rods, does not describe any complications in the lumbo sacral junction.

Broom et al., in a group of 38 patients with fusion to the sacrum , describe 16 with the “windshield-wiper sign”.

In Gau's paper we found 16,1% of complications related to L5-S1 fusion in a group of 68 patients treated with the Luque Galveston technique.

1)Penetration of the pelvic cortex: 2 patients

2) Dislocation from the pelvis: 1 patient

3) Dural tear L5-S1 level: 1 patient,

4) Pseudarthrosis L5-S1:4 patients

5) Broken rod in the lumbosacral area: 3 patients

Sink et al recorded 41 patients with neuromuscular spines that have been fused with the Luque Galveston technique of which 5 (12%) patients have posterior migration of the distal end of the Galveston rods.

In our group of patient 12 were fused to S1. We found 1 complication with protruding tip on the ilium

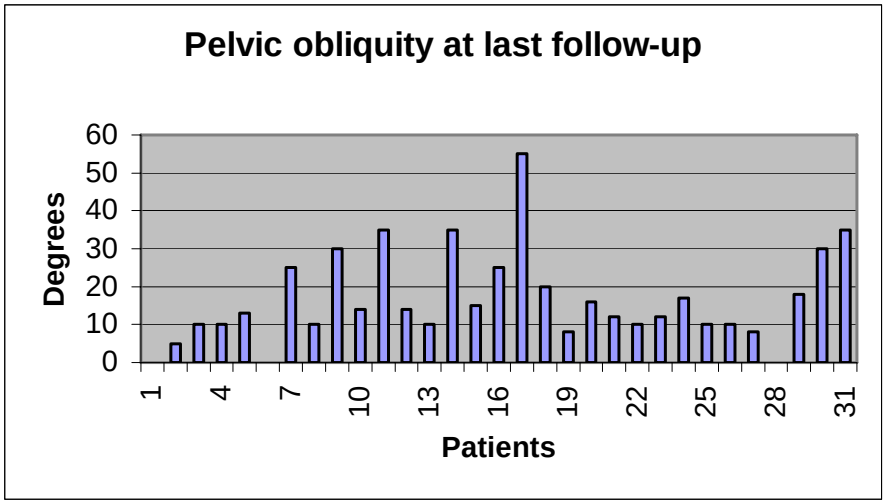
Some authors agree that the indication for spinal fusion to the lumbosacral junction is a pelvic obliquity of more than 10° (39), others say that the lumbar curves have tended to be associated with greater progression of pelvic obliquity. For this reason, they advocate pelvic fixation in curves with an apex at or distal to L1.

In our institution, we consider that the pelvic obliquity can be accepted to a higher degree and that is why we fuse to the sacrum only when the pelvic obliquity is included in the lumbar curve. Pelvic obliquity up to 30° has not been found as problematic for our patients. Only one patient with 25° of pelvic obliquity has pain and a second one with 55° of pelvic obliquity has required several chair adaptations. Otherwise, our study shows 7 patients with a pelvic obliquity between 20 and 35° without complications.

When we evaluated the whole group of 31 patients, the average pelvic obliquity was 16.6°. It is interesting to notice that only the two patients mentioned above were symptomatic (Fig.20).

In agreement with our point of view, Gaine et al. allow up to 30° of pelvic obliquity before fusing to the pelvis. Moreover, Majd and colleagues followed all patients in a nursing home and have found three patients that had decubiti and difficulty in managing pelvic obliquity. All of them had more than 45° of pelvic obliquity.

Fig 20 : Pelvic obliquity at last follow-up. Patient N°7 had lumbosacral pain and Patient N°17 has had multiple chair adaptations (more than once every 2 years) Measurement of pelvic obliquity with the angle between T1-S1 line and the biiliac crest line (33)



When we look at the degree of pelvic obliquity correction from different authors we have found:

-Yasici et al. describe a pelvic obliquity correction of 81% using the Isola Galvestone technique.

- Maloney, with the unit rod, achieved 82% of pelvic obliquity correction with an anterior approach, a scoliosis modification of 52% of the thoracic curve and 62% of the lumbar curve.

-Allen and Ferguson obtain 67% of correction of pelvic obliquity with Galvestone but without an anterior approach.

-Gau et al. showed 55% improvement of scoliosis (73° to 33°) and 53% of pelvic obliquity correction (17° to 8° average correction) with pelvic fixation.

The findings of our study assert that pelvic obliquity was corrected in 11,9% when the fusion was taken to L5, and to 54.4% when fusion was taken to S1.

Comparing the amount of loosening correction of the pelvic obliquity between immediate post operative and last follow-up between both fixations, we observed a 2,6° loosening correction when fusion was taken to L5 and a 3,2° loosening correction when fusion was taken to S1, suggesting that neither improvement nor loosening of the correction of the pelvic obliquity when stopping fusion at L5. Gaine et al found the same results stating in their paper that the fixation of an intrailiac pelvic rod achieved the best immediate correction of the pelvic obliquity, however, fusion to the sacrum or L5 has given a similar long-term correction showing that the fact that the pelvis was not included in the fusion does not mean loosening correction.

Regarding the Duchenne group, the corrections achieved stopping at L5 were maintained over time, demonstrating that when there is no pelvic obliquity at pre-op, the fusion can be limited to L5 and the patient won't subsequently develop a pelvic obliquity.

As we can see in our results, the percentage of correction of pelvic obliquity in the poliomylitis group is similar in L5 and S1 fusion levels. Both of them have more than 50% of pelvic obliquity correction. Moreover, it is important to point out that only in the poliomyelitis group did we achieve more than 50% of correction of pelvic obliquity when fusing to L5.

In our group of cerebral palsy patients, the 3 cases in which fusion was taken to L5 had an average of loosening pelvic obliquity of 5° at last follow-up, which hasn't caused any subsequent problems to the patients for daily life and chair adaptation.

As expected, choosing the right level for fusion is always a challenge. We tried to avoid fusion to S1 because of the loss of motion implied, but never the less all of our 3 cases with some type of ambulation and fusion to S1 , have kept their ability to walk.
This has also been described by Tsirikos et al. who show that out of 24 patients who were fused to S1 and were evaluated 2.86 years post-op there was no alteration in their ambulatory status except for one patient who experienced bilateral hip heterotopic ossification and gradually lost her ability to walk.

When considering functional ability, Lonstein and Akbarnia found greater changes in those patients who were dependant sitters at the pre-operative stage. Post-operatively, there was an improvement in functional ability in more than half of these patients, easier chair adaptation, less difficulty and less time required by personnel in caring for these children. Endurance and the ability to use their hands have been others factors described as improved after surgery. These functional parameters were not taken into account in our research, because they were not included in the pre-operative charts and thus, excluding comparison.
Regarding the level of independence of the different groups with the modified Rancho Los Amigos classification (5), we observed:

L5 group: 12 sitters; 6 home ambulators and 1 community ambulator.

S1 group: 9 sitters, 3 home ambulators.

There have been no changes comparing the level of independence at pre operative and last follow up.

When we looked at the complications related to the pelvic obliquity, Knapp D. and Cortes studied 29 patients who were profoundly mentally retarded and with non ambulatory cerebral palsy, age range between 21 and 52 years (average 34). Within this group, neither pelvic obliquity nor scoliosis or hip dislocation brought about problems related to seating. In this population, wheelchairs were adapted to accommodate even the more severe cases.

This result is not in agreement with Drummond et al. who concluded that the pelvic obliquity should be totally corrected to protect pressure areas associated with unbalanced scoliosis and pelvic obliquity. In their paper, the population included patients without sensation in the buttock area (MMC and spinal cord injury).

In our group of patients, pelvic obliquity was 16° on average, no ulcers were found in relation to the deformity.

When considering chair adaptation related to the pelvic obliquity, of the whole group of 31 patients, only one had problems : a myopathic patient with 55° of pelvic obliquity fused to L5 needing regular chair adaptation to avoid pressure sores.

When considering pain at the lumbosacral area, of the whole group of 31 patients, only one had problems related to the pelvic obliquity : a patient with a spinal muscular atrophy type 2 with lumbosacral pain, with 25° of pelvic obliquity fused to L5. Even though the fusion was taken to L5 the X-ray showed that part of the fusion extended to S1 with a line of pseudoarthrosis. In our opinion, the pain was related to this fact and not to the 25° of pelvic obliquity. Pain medication was enough to manage the problem. No surgery or chair adaptation was needed.

9) Conclusion

Patients with neuromuscular disorders run a great risk of developing spinal deformities and these irregularities are often observed together with a long, sweeping, collapsing curve, pelvic obliquity (60%) and loss of sitting balance.

The indications for surgery include deformity that affects the ability to sit continuously, breathing, eating, comfort, the maintenance of the integrity of the skin surface involved in sitting, pain, visual contact, or a curve that could be predicted-on the basis of the natural history of the disease- to progress to the point that the cardio pulmonary functions would be affected. Other indications for the surgical procedure are curve progression documented on serial radiographs and the magnitude of the curve.

The decision to include the pelvis in the fusion is still debatable.

On, the one hand, we find that the scoliosis correction after fusion either to L5 or to S1 is analogous, but complications when the sacrum is involved tend to be higher.

On the other hand, when S1 is included in the fusion, the degree of correction of the pelvic obliquity rises to 50 to 80%; whereas when fusion is stopped at L5, the correction drops to 10%.

Comparing the amount of loss of correction between the first x-Ray post surgery and last follow-up of the pelvic obliquity between the two types of fixation, a 2,6° loss of correction can be seen when fusion is taken to L5 and a 3,2° loss of correction when fusion is taken to S1. This shows that there has neither been improvement nor loosening correction of the pelvic obliquity when stopping fusion at L5.

In our institution, we consider that some degree of pelvic obliquity is acceptable. Pelvic obliquity up to 30° was not found to be problematic for our patients. Only one patient with 25° of pelvic obliquity suffered from pain and a second one with 55° of pelvic obliquity has required several seating chair adaptations. Otherwise, our study shows 7 patients with a pelvic obliquity between 20 an 35° without complications.

Walking ability was retained in all patients (that had the capacity preoperatively) after surgery.

Our final proposition would be:

Fixation up to S1:

1) The pelvic obliquity is included in the lumbar curve.
2) The pelvic obliquity is more than 25°

Fixation up to L5:

1) The pelvic obliquity is not included in the lumbar curve
2) The pelvic obliquity is less than 25°.

When we fused to L5 we found very small improvement of the pelvic obliquity (up to 2.5°), and as seen in our patients there were no major problems with pelvic obliquity up to 35° we think that 25° can be accepted without problem leaving a good range of safety.

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