Apport de l'imagerie dans l'évaluation des modifications biomécaniques du rachis neuromusculaire fixé par une instrumentation bipolaire

Mathilde Gaume

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Institut de Biomécanique Humaine Georges Charpak – Campus de Paris

THÈSE

présentée par : **Mathilde GAUME**

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Apport de l'imagerie dans l'évaluation des modifications biomécaniques du rachis neuromusculaire fixé par une instrumentation bipolaire

Pr Arnaud DUBORY, Professeur des Universités, HDR, Hôpital Henri Mondor, Paris	Président du jury
M David MITTON, Directeur de recherche, HDR, Université Gustave Eiffel, Lyon	Rapporteur
Pr Wafa SKALLI, Professeur Emérite, HDR, Arts et Métiers Sciences et Technologies, Paris	Directrice de thèse
Dr Lotfi MILADI, Chirurgien orthopédiste, Hôpital Necker Enfants Malades, Paris	Co encadrant
M. Claudio VERGARI, Enseignant-chercheur, HDR, Arts et Métiers Sciences et Technologies, Paris	Co encadrant
Pr Susana QUIJANO-ROY, Professeur des universités, Hôpital Raymond Poincaré, Garches.	Examinatrice
Dr Guillaume RIOUALLON, Chirurgien orthopédiste, Hôpital Saint Joseph, Paris	Examinateur
Mme Pauline ASSEMAT, Chargée de Recherche, Université de Toulouse, Toulouse	Examinatrice
Pr Jean DUBOUSSET, Professeur Emérite, Académie Nationale de Médecine	Invité

THÈSE

Résumé en français

Les scolioses d'origine neuromusculaire sont liées à des anomalies du tonus musculaire, dont le pronostic est aggravé par l'atteinte des muscles axiaux et respiratoires. Le schéma thérapeutique classique consiste en la mise en place d'un traitement orthopédique par corset, suivi par une arthrodèse à l'adolescence. Dans le cas de déformations précoces et rapidement progressives, des techniques chirurgicales d'instrumentation sans greffe permettent de préserver un potentiel de croissance en attente de l'arthrodèse. Cependant, ces techniques comportent un taux élevé de complications, mécaniques et infectieuses, ce qui a amené l'équipe d'orthopédie de l'hôpital Necker à élaborer une technique innovante. Cette technique repose sur une instrumentation sans greffe bipolaire vertébro-pelvienne, réalisée par voie mini invasive. L'objectif principal de ce travail de thèse était d'analyser les résultats post-opératoires à 5 ans de recul chez ces patients. Nous avons remis en question la nécessité d'une arthrodèse vertébrale postérieure en fin de croissance, en raison du maintien de la correction des déformations, après maturation squelettique. Ces constations nous ont fait supposer la survenue d'un enraidissement spontané et progressif du rachis. Nous avons cherché à quantifier cet enraidissement, grâce à différents moyens d'imageries. Sur le plan osseux, la tomodensitométrie a permis de montrer que le montage préservait la croissance de la colonne vertébrale, tout en induisant la fusion de 93 % des articulaires postérieures. L'élastographie discale a apporté des informations complémentaires en montrant une augmentation significative de la rigidité discale. Enfin, l'IRM musculaire a permis de montrer que les muscles spinaux et de la cuisse étaient globalement préservés après la chirurgie, à l'exception du multifidus et du sartorius. En pratique clinique, il nous parait donc justifié de proposer un examen de tomodensitométrie à basse dose après 5 ans de recul, afin de quantifier l'enraidissement vertébral et valider l'absence d'indication de conversion en arthrodèse vertébrale postérieure. L'élastographie discale lombaire pourrait être un outil complémentaire.

Mots-clés : scoliose neuromusculaire, instrumentation bipolaire, enraidissement rachidien.

Résumé en anglais

Neuromuscular scoliosis is related to muscle tone abnormalities, which prognosis is worsened by the involvement of axial and respiratory muscles. The classic treatment consists of a brace, followed by arthrodesis in adolescence. In cases of early and rapidly progressive deformities, fusionless surgery make it possible to preserve a growth potential while waiting for arthrodesis. However, these techniques provide a high rate of mechanical and infectious complications, which led the scoliosis surgeon team at Necker Hospital to develop an innovative technique. This technique is based on a fusionless bipolar fixation from the thoracic spine to the pelvis, performed by a minimally invasive approach. The main objective of this thesis was to analyze the outcomes at 5 years follow-up in these patients. We questioned the need for posterior vertebral arthrodesis at the end of growth, because of the stability of the correction at final follow-up. These findings led us to assume the occurrence of spontaneous and progressive stiffening of the spine. We sought to quantify this stiffening, using different imaging methods. On the bone level, CT scans showed that the construct preserved the growth of the spine, while inducing the fusion of 93% of the facet joints. Discal elastography provided additional information by showing a significant increase in disc stiffness. Finally, muscle MRI showed that the spinal and thigh muscles were globally preserved after surgery, except for the multifidus and sartorius. In clinical practice, it therefore seems justified to us to propose a low-dose CT scan after 5 years of follow-up, to evaluate the spinal stiffening and to validate the absence of indication for conversion to posterior spinal arthrodesis. Lumbar disc elastography could be a complementary useful tool.

Ke-ywords : Neuromuscular Scoliosis, Fusionless Surgery, Bipolar Technique, Minimally Invasive Fusionless Surgery.

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1. Introduction générale

Les maladies neuromusculaires sont liées à des causes musculaires propres, neurologiques centrales ou périphériques, et ce, d'origine primitive ou secondaire (Tableau 1). Leur incidence est très faible, de l'ordre de 2/1000 naissances pour les origines centrales et de 5.1–27.7/100,000 naissances pour l'amyotrophie spinale infantile, qui sont les causes les plus fréquentes[1].

Origine neurologique centrale	Encéphalopathie (Causes pré, péri ou post natales)
Motoneurone central	Ataxie héréditaire (Friedrich) Syringomyélie Autres (encéphalopathie, sd de Rett)
Origine neurologique périphérique	Poliomyélite antérieure aigue
Motoneurone périphérique	Amyotrophie spinale infantile Neuropathie sensitive et motrice héréditaire Neuropathie sensitive et autonome héréditaire (dysautonomie familiale)
Origine mixte (centrale et périphérique)	Lésion médullaire Myélodysplasie Myéloméningocèle
Jonction neuromusculaire	Myasthénie
Origine musculaire	Myopathie de Duchenne Autres dystrophies musculaires Arthrogrypose

Tableau1 : Principales étiologies des scolioses d'origine neuromusculaire

La scoliose neuromusculaire apparait souvent à un âge précoce, dès les premières années de vie, liée à un défaut de soutien du rachis par l'enveloppe musculaire[2,3]. Quand le sujet est jeune, et que le traitement orthopédique par corset n'est plus suffisant pour contenir la déformation, il faut recourir à une prise en charge chirurgicale précoce, qui conserve la croissance vertébrale. Ces techniques d'instrumentations, dites sans greffe, présentent cependant un taux de complications important, rapporté dans la littérature.

Pour améliorer les techniques courantes d'instrumentation sans greffe, le Dr Miladi, chirurgien orthopédiste du rachis à l'hôpital Necker-Enfants malades, a proposé une option innovante, qui repose sur une instrumentation sans greffe bipolaire vertébro-pelvienne, réalisée par voie mini invasive.

Dans ce contexte, l'objectif de cette thèse est d'étudier les modifications biomécaniques à long-terme de la colonne vertébrale chez les patients atteints de scoliose neuromusculaire. Pour cela, nous nous sommes interrogés sur l'évolution clinique et radiologique de ces patients à plus de 5 ans de recul, et nous avons étudié leurs particularités anatomiques après cette chirurgie, sur le plan osseux, discal et musculaire, à l'aide de différents moyens d'imagerie (tomodensitométrie, élastographie discale et IRM musculaire).

2. Rappels anatomiques et cliniques

Le risque d'apparition d'une scoliose dans le cadre d'une maladie neuromusculaire varie de 25 à 100% selon les étiologies[4] (Tableau 2).

Etiologie	Prévalence (en %)
Paralysie cérébrale	25
Myélodysplasie	60
Amyotrophie spinale infantile	67
Ataxie de Friedreich	80
Myopathie de Duchenne	90
Lésion médullaire (<10 ans)	100

Tableau 2 : Prévalence de la scoliose en fonction des étiologies neuromusculaires

Elle est classiquement rapidement évolutive, même après maturité squelettique. Ainsi, chez les patients atteints de paralysie cérébrale, elle peut progresser à l'âge adulte de 1.4° par an pour des courbures principales avec un angle de Cobb>50°[5]. Sa localisation est le plus souvent thoraco-lombaire et s'étend jusqu'au pelvis, entrainant une obliquité pelvienne. L'obliquité pelvienne peut être mixte, d'origine haute, résultant d'une rétraction asymétrique des muscles du tronc et du pelvis, et d'origine basse, en raison d'une asymétrie posturale des hanches, provoquant des rétractions en adduction, flexion et rotation interne. Cette obliquité pelvienne s'associe à une inégalité du poids supporté lors de la position assise à l'origine de douleurs, d'une mauvaise tolérance de la position assise et d'escarres[6]. La scoliose affecte la croissance thoracique et le développement pulmonaire, aboutissant à un syndrome d'insuffisance thoracique, défini par l'incapacité du thorax à assurer une fonction respiratoire et une croissance pulmonaire normale[7]. Pour un angle de Cobb de la courbure principale supérieur à 100°, la capacité vitale fonctionnelle est diminuée de 30 %. Au-delà de 120°, il existe un risque d'insuffisance respiratoire et de cœur pulmonaire chronique[8]. De plus, une hauteur thoracique T1-T12 inférieure à 18-22 cm est associée à une insuffisance respiratoire sévère. La fonction respiratoire est d'autant plus altérée chez les patients atteints de scoliose neuromusculaire, du fait de l'atteinte des muscles respiratoires[9,10].

Le protocole thérapeutique classique est basé dans un premier temps sur un traitement orthopédique, mis en place dès que la déformation rachidienne est constatée, sous la forme d'un corset renouvelé chaque année dès le plus jeune âge, plus ou moins associé à des plâtres correcteurs quand la déformation s'aggrave. Ce traitement orthopédique ne permet souvent pas de corriger la déformation, mais il a pour objectif de la contrôler pendant la croissance en limitant son aggravation[11].

Dans un second temps, le traitement est chirurgical, et consiste en une arthrodèse vertébrale postérieure, réalisée après le pic de croissance, quand le tronc de l'enfant a atteint un degré de maturité osseuse suffisante. Cette technique consiste en une correction de la déformation et sa stabilisation par une instrumentation postérieure, étendue du rachis thoracique haut jusqu'au bassin, associée à un avivement osseux étendu et à une greffe dont le but est d'aboutir à la fusion des vertèbres entre elles sur toute la zone instrumentée.

Cependant, pour les déformations rachidiennes rapidement évolutives, un traitement chirurgical plus précoce, avant l'adolescence, peut être nécessaire.

Ce traitement a longtemps reposé sur l'épiphysiodèse convexe par voie postérieure et/ ou antérieure. Cette chirurgie consiste à réaliser une fusion des corps vertébraux du côté convexe de la courbure, pour en arrêter la croissance, la croissance du côté concave se poursuivant normalement. Cette option chirurgicale permet théoriquement de corriger de manière progressive la courbure scoliotique. Or l'expérience a montré que les résultats de l'épiphysiodèse restent très aléatoires sur le plan de la correction rachidienne, et qu'elle entraine, du fait de la fusion étendue sur plusieurs niveaux, une limitation de la croissance du tronc, du thorax, et une insuffisance respiratoire[12].

A partir des années 1980, les chirurgiens orthopédistes se sont orientés vers un autre type de traitement chirurgical précoce, préservant la croissance de la colonne vertébrale avant la

réalisation d'une arthrodèse à l'adolescence. Il s'agit des techniques d'instrumentations sans greffe dont de multiples types ont été décrits[13,14].

Dans le service de chirurgie orthopédique et traumatologie pédiatrique de l'hôpital Necker, le docteur Miladi a mis au point en 2010 une technique d'instrumentation sans greffe bipolaire que nous allons décrire[15].

3. Description de la technique d'instrumentation sans greffe bipolaire

L'intervention est réalisée sous traction per opératoire modérée. Une traction est appliquée de façon asymétrique en cas de bassin oblique. Dans les cas de scolioses raides, la chirurgie est préparée par une traction de 4 à 6 semaines à l'aide d'un halo crânien. Toute l'intervention se déroule sous monitorage de la conduction médullaire, par analyse des potentiels évoqués somesthésiques et moteurs[16].

La voie d'abord est mini invasive, à travers deux petites incisions médianes, de quelques centimètres chacune, l'une proximale et l'autre distale, en regard des points d'appui, préservant la zone intermédiaire laissée libre.

La fixation proximale est vertébrale, étendue aux cinq premières vertèbres thoraciques (Figure 1). Elle est assurée par une double pince de crochets pédiculo sus-lamaires sur deux vertèbres adjacentes, séparées par une vertèbre libre intermédiaire, de façon bilatérale[17].

La fixation distale est pelvienne, assurée par des vis ilio-sacrées, passant à travers des connecteurs spécifiques, et reliées aux tiges. Les connecteurs sont insérés par voie trans-musculaire paramédiane dans des logettes osseuses, creusées à la face postérieure du sacrum entre l'articulaire L5-S1 et le premier foramen sacré postérieur. Un guide spécifique permet l'introduction des vis ilio-sacrées en percutané à travers l'aile iliaque[18-20].

Ces fixations bipolaires sont reliées de chaque côté par deux tiges télescopiques pré cintrées, de gros calibre (5.5mm), en titane ou en chrome cobalt :

-Une longue tige (en position médiale) descend des crochets proximaux jusqu'à la proximité du connecteur ilio sacré.

- Une tige courte (en position latérale) est fixée au connecteur ilio-sacré.

Elles sont introduites d'une incision à l'autre, et passées en intra musculaire de chaque côté, sans dégagement musculaire ni avivement osseux.

Les deux tiges sont reliées entre elles par deux dominos fermés et se chevauchent sur plusieurs centimètres. La quantité de chevauchement représente la réserve d'allongement du montage, dont la longueur est adaptée au potentiel de croissance résiduel du patient. Elles sont positionnées le long du rachis, le plus parallèlement possible pour faciliter leur coulissement lors des retensions ultérieures.

On procède ensuite à la correction de la scoliose et de l'obliquité pelvienne par des manœuvres de distraction concave et de compression convexe, par l'intermédiaire des dominos, avant de les verrouiller.

Le montage se termine par la mise en place de trois dispositifs de traction transverse : deux dispositifs proximaux sur les grandes tiges, réalisant un montage en cadre, et un dispositif distal sur les petites tiges, stabilisant les vis ilio-sacrées.

Les ré- interventions de retension de tiges sont réalisées à travers l'incision distale tous les 12 à 24 mois, permettant soit de suivre la croissance rachidienne, soit de poursuivre la correction des déformations résiduelles, notamment pour les bassins obliques.

Actuellement, de nouvelles tige auto-extensibles NEMOST®, selon le même principe bipolaire permettent d'éviter les interventions répétées de retention de tiges [21].

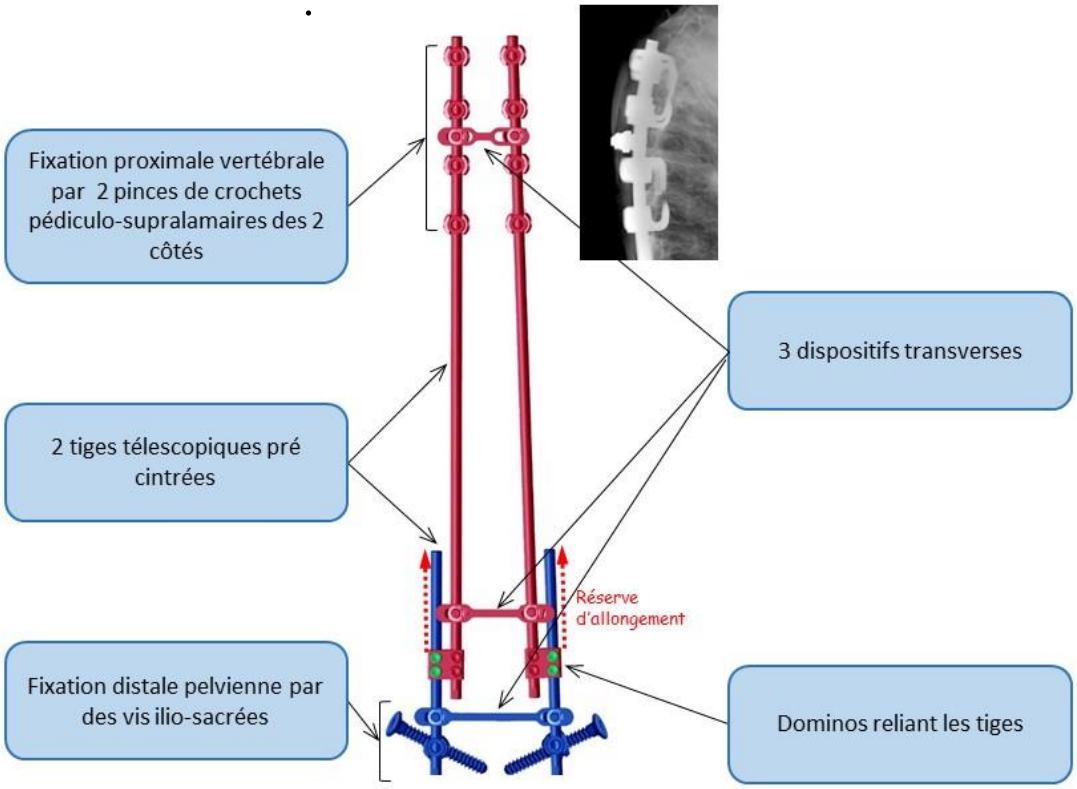

Figure 1 : Schéma de synthèse du montage d'instrumentation sans greffe bipolaire

4. Revue de la littérature

4.1. Les techniques d'instrumentation sans greffe pour le traitement des scolioses neuromusculaire sévères de l'enfant

De nombreux types d'instrumentations sans greffe ont été décrites, dont le tableau 3 résume les principaux types et résultats, avec un taux de complications de l'ordre de 40 à 73% selon la littérature.

Auteurs	Technique	Patients	Etiologie	Recul	Angle de Cobb (% correction)	Obliquité pelvienne (% correction)	%Complications globales
Ouellet et al.[22]	Luque trolley	15	Mixte	2 ans	61	-	40% dont : -100% mécanique
Mc Carthy et al.[23]	Shilla	38	Mixte	7 ans	55	-	73% dont : -63% mécanique -15% infection
Abol et al.[24]	VEPTR	20	Neuro Musculaire	< 2ans	24	43	45% dont : -60% mécanique -15% infection
Akbarnia et al.[25]	Tige de croissance standard	23	Mixte	2 ans	56	-	47%
Sponseller et al.[26]	Tige de croissance standard	36	Neuro Musculaire	3 ans	55	40	44% dont : -30% mécaniques -16% infections
Miladi et al.[15]	Tiges de croissance bipolaires	100	Neuromusculaire	2 ans	61	83	26% dont : -12%mécaniques -16% infections

Tableau 3 : Taux de correction et complications obtenues avec les instrumentations sans greffe

Les raisons principales des complications sont d'ordre :

- Mécaniques : favorisées par l'ostéoporose de ces patients dénutris, qui ne permet pas de supporter les contraintes, notamment les ancrages costaux et les ancrages pelviens instables. L'utilisation de tiges de trop petit diamètre (4.5 mm ou moins) contribue aussi à la survenue de complications mécaniques.

- Biologiques : une chirurgie étendue, avec un large abord sous périosté et une dissection de la zone intermédiaire, favorise la survenue de la fibrose et des ossifications spontanées.

- Stratégiques : une construction trop rigide, des tiges obliques ou mal cintrées, ainsi que des erreurs dans le choix et dans le nombre des vertèbres porteuses.

Si les résultats de la cohorte historique des 100 premiers patients de la série de Miladi et al. sont prometteurs à deux ans de recul, un des objectifs de la thèse était d'explorer les résultats de cette technique à long-terme, à plus de cinq ans de recul. Nous nous sommes également interrogés sur les options thérapeutiques envisageables à la fin de la croissance.

4.2. Options thérapeutiques à la fin de la croissance

A maturité squelettique, une fusion finale par arthrodèse vertébrale postérieure est traditionnellement proposée par les chirurgiens. L'arthrodèse vertébrale postérieure est une intervention lourde, associée à un taux de complication élevé, notamment hémorragique dans cette population fragile de patients. Poe-Kochert et al[27]. ont constaté sur une série de 100 patients, initialement traités avec des tiges de croissance standards puis arthrodèses, que 20% d'entre eux ont présenté 30 complications, nécessitant une réintervention (57 procédures), avec en moyenne 1.5 complication/patient dans les deux ans suivant l'arthrodèse.

Seuls deux auteurs évoquent dans la littérature la possibilité de se dispenser d'une arthrodèse vertébrale postérieure finale après instrumentation sans greffe.

Amit Jain et al.[28]proposent de conserver le montage à la fin de croissance, si les patients ont obtenu une correction de la déformation et une hauteur de tronc satisfaisants, et qu'il n'y a pas de preuve clinique ou radiographique de complications liées aux implants.

C'est le cas également de Bouthors et al.[29,30], dans une série de patients atteints de neurofibromatose de type 1, dont aucun des 18 patients n'a été converti en arthrodèse vertébrale postérieure.

Cependant, la décision de maintenir le montage initial repose chez ces auteurs sur des observations cliniques. Nous faisons l'hypothèse de la survenue d'un enraidissement rachidien progressif après instrumentation sans greffe, qui permettrait de rendre inutile une conversion en arthrodèse vertébrale postérieure.

Nous avons cherché à caractériser la survenue d'un enraidissement rachidien à l'aide de différents moyens d'imageries. Tout d'abord, nous nous sommes intéressés aux structures ostéo-articulaires.

4.3. Outils d'imagerie à disposition pour l'évaluation de l'enraidissement rachidien chez les patients atteints de scoliose neuromusculaire

4.3.1. La tomodensitométrie

La tomodensitométrie est l'examen de référence pour étudier les structures osseuses. Elle a été utilisée dans plusieurs études qui ont quantifié la fusion osseuse après chirurgie rachidienne chez l'adulte[31].

Auteurs	Technique	Critère d'évaluation
Peters et al.[32]	Arthrodèse vertébrale lombaire	Nombre de ponts osseux dans le plan coronal : 0 : Pas de pont 1 : Sur un côté à l'intérieur ou autour d'une cage, à droite ou à gauche 2 : Sur les deux côtés à l'intérieur ou autour des cages
Shah et al.[33]	Cages intersomatiques en titane et instrumentation transpédiculaire	Présence de ponts osseux entre cages intervertébrales -Sur les vues coronales et sagittales -Latéral / médial / ant / post -Oui/non
Berjano et al.[34]	T-LIF lombaire (Transforaminal Lumbar Interbody Fusion)	-Complètement fusionné : Pont osseux présent. -Probablement fusionné : Radio-transparence partielle -Pseudarthrose
Carreon et al.[35]	Arthrodèse vertébrale lombaire	-Fusion des articulations postérieures (Les deux / une/ aucune) -Fusion des gouttières postérolatérales (Les deux/ une / aucune)

Tableau 4 : Evaluation de l'autofusion vertébrale par tomodensitométrie, après chirurgie rachidienne.

Néanmoins, aucune étude à notre connaissance n'a évalué les modifications osseuses des scolioses neuromusculaires opérées ou non, par instrumentation sans greffe. Ceci est expliqué par le caractère irradiant de la tomodensitométrie, qui en fait un outil peu utilisé en routine dans la population pédiatrique.

Par ailleurs, la tomodensitométrie n'est pas un examen de choix pour l'évaluation des structures discales après instrumentation.

4.3.2. L'élastographie discale

L'étude des structures discales nous parait importante, car les disques vertébraux assurent la souplesse de la colonne vertébrale. Certains auteurs ont cherché à étudier l'enraidissement des disques vertébraux induit par la chirurgie rachidienne en utilisant l'élastographie. Il s'agit d'une technique d'imagerie basée sur la vitesse de propagation de l'onde de cisaillement induite par les ultrasons, pour quantifier l'élasticité ou la rigidité des tissus mous[36]. En pratique, elle associe une sonde permettant de palper le disque vertébral à l'aide d'une onde focalisée, et un échographe ultra-rapide qui enregistre la propagation de cette onde au rythme de 1 000 images à la seconde. Récemment, Langlais et al.[37] ont montré que les patients atteints de scoliose idiopathique présentaient un anneau fibreux plus rigide que les adolescents asymptomatiques, avec des vitesses d'ondes de cisaillement de 3,5 SD 0,3 m/s contre 3,0 SD 0,3 m/s, respectivement.

L'utilisation de cette technique n'a jamais été utilisée pour les scolioses neuromusculaires. Elle a l'avantage d'être de réalisation rapide et non invasive, raison pour laquelle nous l'avons retenue pour évaluer la rigidité des disques vertébraux lombaires après instrumentation sans greffe bipolaire.

4.3.3. L'IRM musculaire

Nous avons enfin cherché à caractériser les modifications structurales musculaires. Pour cela, nous avons choisi l'IRM musculaire, qui est une technique non invasive, non irradiante, et de plus en plus utilisée pour établir un diagnostic, caractériser la gravité et surveiller la progression des maladies neuromusculaires[38,39].

De nombreuses études ont évalué les modifications musculaires post-opératoires après chirurgie rachidienne dégénérative de l'adulte (Tableau 4). Ces dernières évaluent essentiellement l'aire de section corporelle (ou CSA cross-sectional area, en cm²) afin d'étudier les lésions musculaires para vertébrales en fonction de différentes voies d'abord ou de techniques d'instrumentation, ou pour évaluer le retentissement sur les douleurs ou la qualité de vie.

Auteurs	**Objectifs**	**Patients**	**Age moyen**	**Séquences**	**Analyse musculaire**
Teraguchi et al.[40]	Relation entre les zones de haute intensité et lombalgie/sciatique	1214	48 ans	T2	Classification des zones de haute intensité
Tandon et al.[41]	Corrélation entre les lésions musculaires et le score d'Oswestry	30	>18 ans	T1	Multifidus CSA (cm²)
Gille et al.[42]	Evaluation des modifications de l'erector spinae après chirurgie lombaire par une voie postérieure	20	48 ans	T1	Erector spinae CSA (cm²)
Kang et al.[43]	Comparaison de la musculature paraspinale chez des patients atteints de cyphose dégénérative lombaire et de lombalgie chronique	108	44 à 74 ans	T1, T2	Psoas, erector spinae, multifidus CSA (cm²)
Motosuneya et al.[44]	Comparaison de l'atrophie musculature rachidienne entre 5 groupes chirurgicaux.	49	>36 ans	T2	Ratio d'atrophie (pré/post CSA) (cm²)

Tableau 5 : Revue de la littérature utilisant l'IRM pour l'analyse des modifications musculaires post opératoires. CSA : cross-sectional area

Dans la population de patients atteints de pathologies d'origine neuromusculaire, aucune étude à notre connaissance n'a été réalisée après chirurgie rachidienne. L'essentiel de la bibliographie repose sur des mesures qualitatives de muscles des membres inférieurs (Tableau 5).

Auteurs	Etiologie	Age moyen	Localisation	Séquences IRM	Analyse musculaire
Brogna et al.[45]	ASI II et III	2 à 45 ans	MI	T1	Remplacement graisseux selon Mercuri (1 : normal, 2 : minime, remplacement graisseux < 30%, 3 : modérée remplacement graisseux 30-60%, 4 : sévère remplacement graisseux > 60%)
Sproule et al.[46]	ASI I, II et III	5 à 47 ans	MI	T1	-Volumes musculaires avec signal relativement "normal" ou "anormal". -Volume musculaire (mL/cm)
Ueno et al.[47]	ASI II	Case reports 5 et 2 ans	MI	T1, T2 fat sat	-Présence d'une atrophie musculaire (oui ou non) -Présence d'une infiltration graisseuse (oui ou non)
Liu et al.[48]	ASI II et III	5 à 17ans	MI	T1	-Augmentation de la graisse sous-cutanée (oui ou non) -Infiltration graisseuse (oui ou non)

Tableau 6 : Méthodes utilisées pour la caractérisation de la trophicité musculaire chez les patients atteints d'amyotrophies spinales infantiles de type II et III. ASI : Amyotrophie spinale infantile ; MI : membres inférieurs.

Cette revue de la littérature nous a permis de voir qu'il existe de nombreuses techniques d'instrumentation sans greffe pour le traitement des scolioses neuromusculaire sévères de l'enfant, et que la conduite à tenir à la fin du programme de croissance n'est pas consensuelle.

Nous avons vu que les résultats des différentes instrumentations sont limités pour la plupart des études à deux ans, il nous parait indispensable d'analyser leur devenir à plus long terme. Cela fera l'objet de la première partie du travail.

Nous supposons qu'un enraidissement vertébral progressif s'instaure, et qu'il pourrait justifier d'éviter une arthrodèse vertébrale postérieure à maturité squelettique.

La deuxième partie abordera les différents outils d'imagerie à disposition pour explorer cet enraidissement rachidien : les structures osseuses à l'aide de la tomodensitométrie, les disques vertébraux à l'aide de l'élastographie, et les structures musculaires à l'aide de l'IRM musculaire. Nous présenterons les travaux réalisés dans cet objectif.

5. Résultats cliniques et radiologiques à long terme de l’instrumentation sans greffe bipolaire

Forts de cette revue de littérature, nous nous sommes intéressés dans un premier temps à l’évaluation des résultats à long terme de l’instrumentation sans greffe bipolaire, sur le plan clinique et radiologique, afin de confirmer la stabilité de la correction des déformations obtenue par cette technique, à plus de cinq ans de recul. C’est ce qui a fait l’objet de l’article ci-après, publié dans la revue *Spine.* Il s’agit d’une étude rétrospective concernant les cent premiers patients opérés selon cette technique.

5.1. Article 1 : Minimally Invasive Surgery for Neuromuscular Scoliosis: Results and Complications at a Minimal Follow-up of 5 Years.

Gaume M, Vergari C, Khouri N, Skalli I W, Glorion C, Miladi L.

***Spine* (Phila Pa 1976). 2021 Dec 15 ;46(24) :1696-1704.**

Résumé

Introduction : Le traitement conservateur n'est pas efficace dans les scolioses neuromusculaires progressives. La chirurgie précoce à l'aide de tiges de croissance (TC) est de plus en plus préconisée pour contrôler la déformation tout en préservant la croissance rachidienne et thoracique avant arthrodèse vertébrale postérieure. Cependant, ces techniques présentent encore un taux élevé de complications.

Objectif : L'objectif de cette étude était de rapporter les résultats d'une technique innovante, consistant en une instrumentation sans greffe par voie mini invasive, avec recul minimum de 5 ans.

Méthodes : La technique repose sur une instrumentation bipolaire avec une double tige télescopique, ancrée par une double pince de crochets en proximal, et par des vis ilio-sacrées en distal. Les résultats cliniques et radiologiques des premiers 100 patients consécutifs atteints de scoliose neuromusculaire, et qui ont subi cette chirurgie avec un suivi minimum de 5 ans ont été évalués.

Résultats : 6,5 ± 0,7 ans après la chirurgie initiale, 6 patients ont été perdus de vue et 11 sont décédés pour des raisons indépendantes. Sur les 83 patients restants au dernier suivi, l'angle de Cobb moyen était stable à 35,0°, ce qui correspond à une correction de 61 % de la déformation initiale. L'obliquité pelvienne moyenne était de 29,6° (0,3° à 80,0°) en préopératoire et de 7,2° (0,2° à 23,5°) au dernier suivi. La correction de l'hypercyphose est restée stable. La maturité squelettique a été atteinte chez 42/83 patients (50,6%). Aucun de ces patients n'a eu besoin d'une arthrodèse vertébrale postérieure. Le taux global de complications était de 31,3 %.

Conclusion : Les résultats à 5 ans de recul de cette technique mini-invasive sans greffe ont montré une correction stable des déformations rachidiennes et de l'obliquité pelvienne, avec un taux de complication réduit. Une arthrodèse vertébrale postérieure n'a pas été nécessaire pour tous les patients à maturité squelettique. Cette technique pourrait donc devenir une alternative à l'arthrodèse pour les scolioses neuromusculaires.

Abstract

Study Design: A prospective study

Summary of background data: Conservative treatment is not effective in progressive neuromuscular scoliosis. Early surgery using growing rods (GR) are increasingly advocated to control the deformity while preserving spinal and thoracic growth before arthrodesis. These techniques still provide a high rate of complications.

Objective: The aim was to report the results of an alternative technique to GR for neuromuscular scoliosis using a minimally invasive fusionless surgery with a minimum of 5 years follow-up.

Methods: The technique relies on a bilateral double rod sliding instrumentation anchored proximally by 4 hooks claws and distally to the pelvis by ilio-sacral screws through a minimally invasive approach. The clinical and radiological outcomes of 100 consecutive patients with neuromuscular scoliosis who underwent this fusionless surgery with a minimum follow-up of 5 years were reviewed.

Results: 6.5 ± 0.7 years after initial surgery, 6 patients were lost of follow-up and 11 died of unrelated raison. Of the 83 remaining patients at latest follow-up, mean Cobb angle was stable to 35.0° which correspond to 61% correction of the initial deformation. Mean pelvic obliquity was 29.6° (0.3° to 80.0°) preoperatively and 7.2 (0.2° to 23.5°) at latest follow-up. Correction of the hyper kyphosis remained stable. Skeletal maturity was reached in 42/ 83 patients (50,6%). None of these patients has required spinal fusion. The global complication rate was 31.3%.

Conclusion: The outcomes of this minimally invasive fusionless technique at 5 years follow-up showed a stable correction of spinal deformities and pelvic obliquity over time, with a reduced rate of complication. The arthrodesis was not required for all patient at skeletal maturity. This technique could be a good alternative to arthrodesis for neuromuscular scoliosis.

Level of evidence: 3

INTRODUCTION

Neuromuscular scoliosis is caused by a disorder of the brain, spinal cord or muscular system. Neuromuscular curves are often associated with trunk imbalance, pelvic obliquity and kyphosis, starting to develop at an early age and usually worsen over time despite conservative treatment [1]. Common conditions associated with neuromuscular scoliosis are cerebral palsy, myelomeningocele, spinal muscular atrophy and muscular dystrophy. Early posterior spine

fusion (PSF) used to be performed for the most severe of them, with the risk of stunting trunk growth and concomitant effects on lung development. Arthrodesis provides a high complication rate, especially infections and hemorrhages requiring long intensive care unit stay [2,3]. Growth-sparing surgical procedures have therefore been developed in order to preserve spinal and thoracic growth and to postpone arthrodesis [4,5]. These techniques were first introduced by Harrington in 1962 [6]. They became more and more popular during the two last decades with the advent of traditional growing rods (TGR). After initial surgery, some surgical procedures of rod lengthening are performed to achieve as spinal height as possible, followed by a PSF when the patient reaches skeletal maturity [7, 8]. However, the complication rate can reach 40 to 73% [9-12] with these methods.

An original technique of bipolar instrumentation was developed as an alternative to TGR for the treatment of neuromuscular scoliosis [13]. The surgical approach was minimally invasive, since proximal and distal short incisions were performed thus preserving the intermediate area to avoid early fibrosis and auto-fusion. The results were maintained after 2-years follow-up with a 61% correction of the Cobb angle and 83% correction of the pelvic obliquity. The aim of this work was to evaluate the clinical and radiological outcomes of minimally invasive fusionless technique for neuromuscular scoliosis at a minimum of 5years follow-up.

PATIENTS AND METHODS

Study design and data collection

Medical charts of the first 100 consecutive patients who underwent surgery for neuromuscular scoliosis at our department between 2011 to 2015 were reviewed. The study was approved by the appropriate ethics committee (CPP, ID-EUDRACT #2014-A01043-44).

Informed consent was obtained from parents/guardians of all participants. The original minimally invasive bipolar technique used in these patients has been previously described [13]. Patients were operated under intra operative traction and evoked potential monitoring. Two short incisions, one thoracic and one lumbar were made to insert the anchors. The instrumentation extends from T1 to the sacrum, proximally with a double pediculo-supralaminar claw and distally why ilio-sacral screws. Two long pre-curved rods (diameter 5.5mm) are inserted in an intramuscular way from the proximal wound to the distal one and attached proximally to the hooks and distally to two short rods fixed inside the ilio-sacral connectors. Side-to-side connectors and crosslinks connect the rods to build a solid sliding frame construct.

In this technique, the correction is obtained through distraction maneuvers that are applied at the distal part of the sliding construct, without further rod manipulations. The bipolar concept relies on a continuous tension between the proximal and the distal points of fixation obtained with rod lengthening on demand. Rods were lengthened using the previous distal incision to access the side-to-side connectors. Lengthening procedure was performed according to the following indications:

- to perfect pelvic correction in case of major residual pelvic obliquity, in particular thanks to the possibility of performing asymmetrical lengthening of the rods;

- to improve a persistent trunk imbalance in the frontal or sagittal plane.

The amount of rod lengthening was from15 to 30mm.

For each patient, the following radiographic parameters were recorded before surgery, immediately after surgery, and at last follow-up: stages of skeletal maturity as reflected by the Risser sign, Cobb angle, pelvic obliquity, and thoracic kyphosis. Screw loosening was defined as a radiolucent area (≥1 mm in circumference) around the screw, noted on plain radiograph by at least two observers. Radiographic measurements were performed using previously described

methods [14,15]. All types of complications were reviewed including implant failure (e.g., malposition of the ilio-sacral screw, hooks migration, and implant breakage) and infections. Mean body weight was recorded preoperatively and at latest follow-up.

Statistical analysis

Variables were described as mean (range). Changes in radiological parameter values from baseline to last follow-up were evaluated by applying the pairwise Student's *t* test. *P* values below 0.05 were considered significant. Complications were described as n (%).

RESULTS

Demography

Of the first 100 patients, 6 were lost of follow-up and 11 died of independent causes from spinal surgery. As such, 83 patients (83% of the initial number of patients) were included (40 Females, 43 Males). All patients were Risser stage 0 at initial surgery with a mean age of 11.5 years (5.2 to 15.4). The diagnoses were cerebral palsy (n=45), spinal muscular atrophy (n=21), muscular dystrophy (n=10), and other neuromuscular disorders (n=7). Mean follow-up was 6.5 years (5.3 to 9.0). 13 patients were ambulatory. Mean body weight was 24.4±6 kg preoperatively and 37.3±7 kg at latest follow-up. (Table 1)

Loss of follow-up, n Unrelated death	6/100 11/100
Follow-up duration, years, mean (range)	6.5 (5.3 to 9.0)
Age at surgery, years, mean (range)	11.5 (5.2 to 15.4)
Gender (F/M)	40/43
Diagnosis, n Cerebral palsy Spinal muscular atrophy Muscular dystrophy Other neurological conditions	 45 21 10 7
Non-ambulatory/ambulatory, n	70/13
Body weight (kg) Preoperatively At latest follow-up	 24.4±6 37.3±7
Rod lengthening per patients, mean Interval between initial surgery and 1st lengthening, years, mean Interval between lengthenings, years, mean Interval between final lengthening and initial surgery, years, mean	1.3 (0 to 4) 1.6±0.6 1.3±0.6 2.8±1.0

Table 1: Main features of the 100 patients treated with fusionless surgery for neuromuscular scoliosis.

The average operative time for the initial surgery was 2 hours 26 minutes (from 1h 33min to 3h 39min; the average operative time for rod lengthening was 44minutes (from 26min to 1h 10min.

24% of patients required 1 to 2 blood transfusion at initial surgery, none after rod lengthening.

Radiological outcomes

Table 2 and 3 reports the changes in these radiological parameters.

	Preoperative	Postoperative	Last follow-up	*P* value
Cobb angle, °, mean (range)	89.0° (46.1° to 149.4°)	33.0° (5.0° to 65.8°)	35.0° (6.4° to 68.5°)	0.33
Pelvic obliquity, °, mean (range)	29.6° (0.3° to 80.0°)	12.2° (0.1° to 35.2°)	7.2° (0.2° to 23.5°)	0.28
Hyper kyphosis T4-T12 >50° mean (range)	69.3° (51.8° to 102.1°)	32.0° (13.1° to 52.1°)	31.3° (8.8° to 50°)	0.31
T1-S1 length, mean, cm	30.02 cm (22.30– 40.13 cm)		36.0cm (29.3-44.2cm)	

Table 2: Radiographic data of the 83 patients

Patients at skeletal maturity (Risser>4), n, %	42/83 (50,6%)
Interval between initial surgery and skeletal maturity, years, mean.	2.9±1.4y
Interval between skeletal maturity and latest follow-up, years, mean.	3.7±1.1
Cobb angle, °, mean (range) at last follow-up.	33.9° (6.4° to 53.4)
Pelvic obliquity, °, mean (range) at last follow-up.	6.9° (0 to 19.9°)
Hyper kyphosis T4-T12 >50° mean (range) at last follow-up	23.2° (14.3 to 37.2°)

Table 3: Radiographic data of patients at skeletal maturity

17 patients did not require rod lengthening thanks to a stable and satisfying correction after initial surgery. The other patients underwent one rod lengthening (30), two rod lengthening (24), three rod lengthening (11) or maximum four rod lengthening (1) to achieve progressive correction of spinal deformity and/or pelvic obliquity.

Screw loosening was visible radiographically in 17 patients (7 ambulatory and 10 non-ambulatory) (Figure 1) No clinical or mechanical consequences of osteolysis were seen in any patients, and no revision was needed.

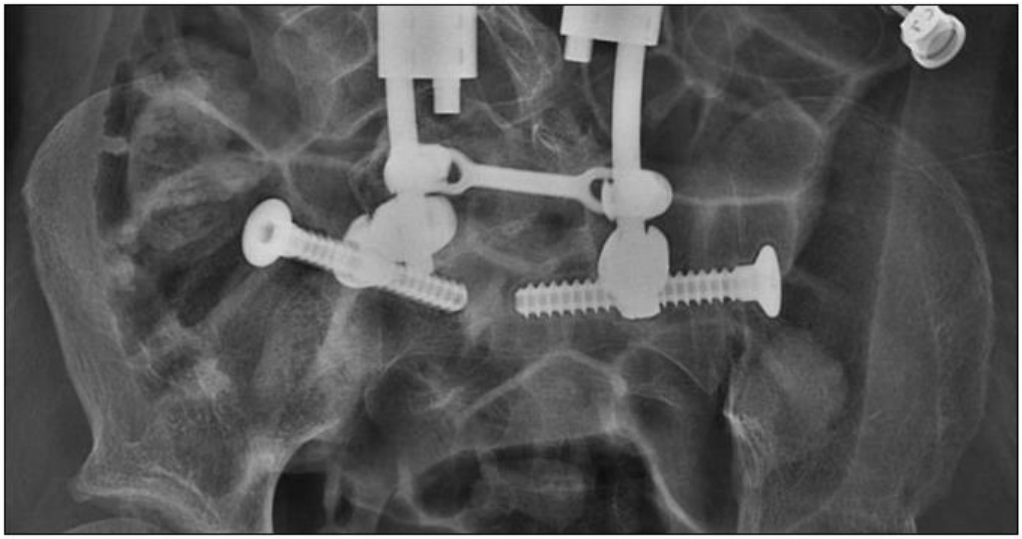

Figure 1 : Screw loosening around ilio-sacral screw

At last follow-up, skeletal maturity (Risser stage 4) was reached in 42/ 83 patients (50.6%) with a mean of 2.9±1.4y after initial surgery. Mean Cobb angle was 34.5° (min 6.4 to max 51.7°) and mean pelvic obliquity was 7.4° (min 0° to max 20.3°). At last follow-up, none of these patients has required PSF with a mean interval of 3.7±1y after skeletal maturity.

Complications

27 complications (12 mechanical and 15 infections) occurred in 26/83 patients corresponding to a global rate of 31%. Table 4 details the 12 mechanical complications and 15 infections requiring unplanned surgery.

Mechanical complications (n=12)	
Proximal hook dislodgment	3
Malposition of ilio-sacral screw	4
Rod breakage Cerebral palsy Ambulatory/ non ambulatory status Mean body weight	5 5/5 1/4 43.3±6 kg
Infectious complications (n=15)	
Diagnosis Cerebral palsy Spinal muscular atrophy Muscular dystrophy	 11 3 1
Mean body weight (kg) Preoperative Last follow -up	 21.7±4 *(p>0.05)* 35.1±6 *(p>0.05)*
Age at initial surgery	10.7±2 *(p>0.05)*
Delay Within the first 3 months of the initial surgery After rod lengthening procedures	 3 12
Management Irrigation and debridement, with appropriate antibiotic therapy Implant removal followed by a new instrumentation Implant removal without new instrumentation	 11 2 2

Table 4: Complications with unplanned surgery of the minimally invasive surgery for neuromuscular scoliosis (n=27, in 26/83 patients)

Mechanical complications

Mechanical complications included 3 cases of proximal hook migration, 4 cases related to the malposition of ilio-sacral screw, and 5 cases of rod breakages.

Rod breakages occurred in a mean interval of 4.0±1 y after initial surgery and were located at lumbar area. Of these patients, pattern curves were thoracolumbar left (3), thoracolumbar right (1) and double major (1) with no significant difference compared with the rest of the cohort

(p>0.05). All patients suffered from cerebral palsy with severe dystonia or hyperactivity. Mean body weight of these 5 patients was 43.3±6 kg (p=0.04) and one of them was a walking patient.

Infectious complications

Of the 15 wound infections, 3 occurred within the first 3 months of the initial surgery and 12 after rod lengthening procedures. 11 were successfully treated by irrigation and debridement, with appropriate antibiotic therapy. 4 patients required implant removal because of chronic infection: 2 of them necessitated a new instrumentation 1 year later because of a deformity worsening; the 2 others did not need new implantation thanks to a stable correction of the deformity. There was significant difference related to mean body weight and age at initial surgery compared with the other patients of the cohort. Most of patients (11/15) suffered from cerebral palsy with a significant difference (p=0.039).

DISCUSSION

In this study, the outcomes and complications of bipolar minimally invasive surgery in children with neuromuscular scoliosis were reviewed with a mean follow-up of 6.5 years. Results showed stable correction of spinal deformity and pelvic obliquity over time, including patients at skeletal maturity. PSF was not required for any patient at last follow-up.

This technique was initially used as an alternative to TGR to reduce mechanical and infectious complications while waiting for PSF planned at skeletal maturity for all of them. However, stable clinical and radiological evolution were noticed with a reduced rate of mechanical complications that did not require performing PSF as initially planned while obtaining an acceptable growth of the spine. Moreover, fibrosis and spontaneous fusion around the implants and along the rods was observed during revision surgeries for rod lengthening. (Figure 2).

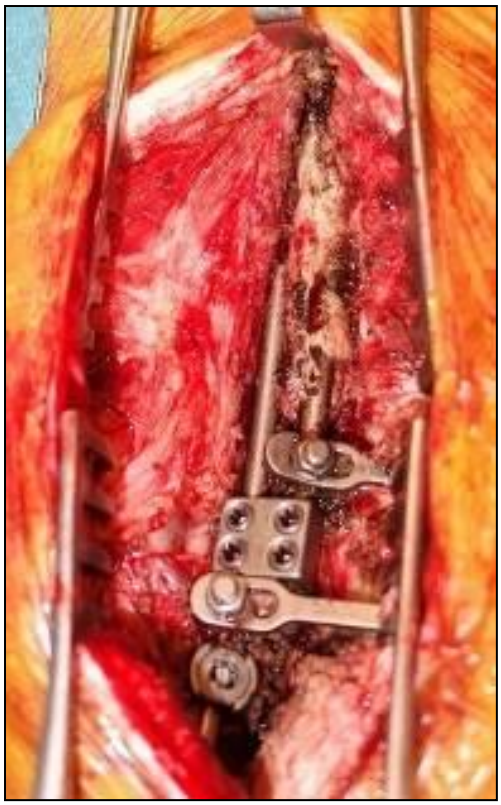

Figure 2 : Fibrosis and spontaneous fusion observed around the implants during a rod lenghtening procedure.

This delayed spontaneous fusion was asserted by lateral bending X-rays performed after hardware removal because of chronic infection, in one patient 8 years after initial surgery. This dynamic X-rays showed the total stiffness of the spine completely fused (Figure 3).

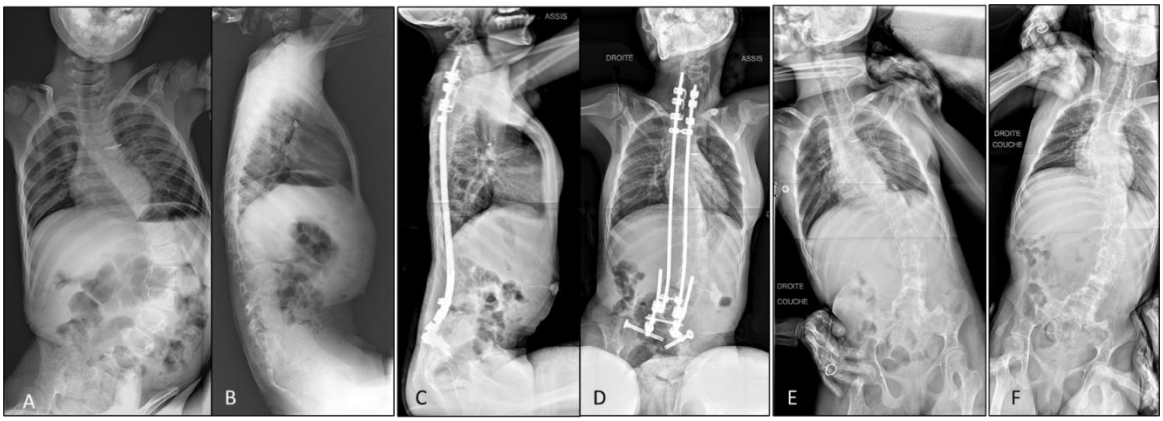

Figure 3 : A 15years old girl with cerebral palsy.A/B, Initial AP and lateral X-Rays. C/D, After initial surgery. E/F, Lateral bending after removal of instrumentation for chronic infection, 8 years after initial surgery.

These observations lead to hypothesize that a progressive stiffening of the spine occurs over time under the influence of the permanent presence of rigid metallic rods. However, this concept of auto-fusion is not new. In 1984, Moe et al. [16] noted spontaneous fusion at the curve extremities and/or at the curve apex in young children treated for severe deformities with subcutaneous Harrington rod without fusion. Mardjetko et al. [17] observed extensive fibrosis and laminar spontaneous fusions under the instrumented regions with Luqué rods, frequently involving the entire thoracic spine but also extending across the thoraco-lumbar spine. In Cahill et al [18] series, the rate of auto-fusion in children treated with growing rods was 89%. This auto-fusion was characterized by solid sheet of bone similar to a mature fusion mass and lead to only moderate correction at the time of definitive fusion. Jain et al. [19] also noticed a progressive ankylosis after GR surgeries and suggested to avoid PSF at skeletal maturity for patients with satisfactory final alignment and trunk height, and no clinical or radiographic evidence of implant-related problems. Recently, Bouthors et al. [20] considered retaining dual growing rods instead of PSF in patients at skeletal maturity with satisfactory deformity correction.

The auto-fusion in traditional growing rods occurs at early stage due to the damage of the intermediate area secondary to repetitive rod lengthening procedures. This auto-fusion is unwanted and participates to the "law of diminishing return" [21]. That is why rod lengthening procedure were recommended with an interval of 6 months. The PSF realized at the end of traditional growing rods series with large auto-fusion was required because the deformity correction was unsatisfactory. In these cases, PSF were frequently performed earlier, before skeletal maturity.

In the bipolar minimally invasive technique, the auto-fusion is expected. It occurs at a delayed stage thanks to the preservation of the intermediate area between the two bipolar fixations and also thanks to the permanent presence of metallic rods. This phenomenon was experienced with

Luqué-Trolley technique in the seventies in which rods were sliding despite spontaneous fusion [22,23].

The avoidance of PSF is particularly beneficial in this fragile neuromuscular population characterized by medical comorbidities, impaired bone quality and poor nutritional status. Compared with other types of scoliosis, complication rates of PSF procedures in neuromuscular scoliosis shows the highest rate of complications (24 to 75%), followed by congenital (10.6%) and idiopathic (6.3%) scoliosis [24]. More specifically, Connie Poe-Kochert et al. [25] reported a mean of 1.5 complications per patient after PSF following treatment with the use of growing rods.

Unlike others growing rods techniques that are mainly provisory to wait for arthrodesis at skeletal maturity, this bipolar technique may be considered as an alternative to arthrodesis as it is strong enough to be definitive. The avoidance of PSF in our series relies on two main advantages of the instrumentation.

The first one is the relatively large deformity corrections of the Cobb angle (61%) and the pelvic obliquity (83%) which remained stable at latest follow-up (Figure 4-5). These amounts of correction were similar or better to TGR series [25,26] and even PSF [27-31].

Figure 4 A/D: Radiographs of a boy with cerebral palsy operated at 7 years old. A/B, Preoperative X-rays. /D X-rays after after 8 years follow-up.

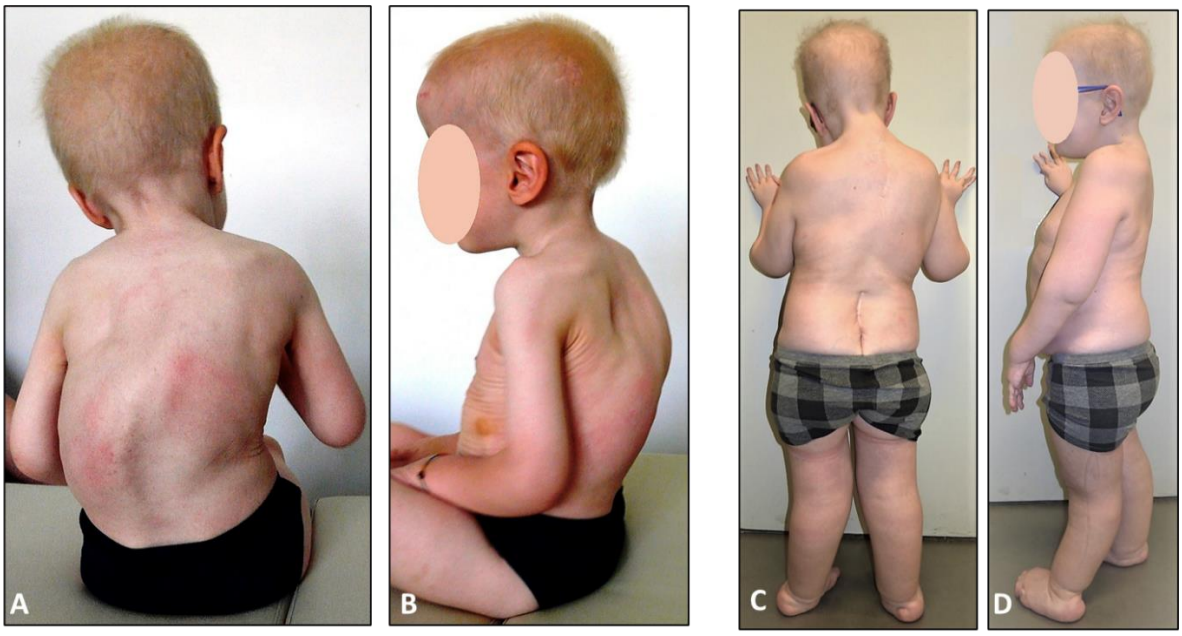

Figure 5 A/D: Pre- and latest photographs after 8 years follow-up of the patient treated by the minimally invasive fusionless surgery.

The second one is the strength and the stability of this type of fixation over time allowing PSF avoidance with a reduced mechanical complication rate. The bipolar construct differs from TGR by the stability of the proximal and distal fixations, the frame sliding construct and the absence of intermediate area damage. Sponseller et al. [9] reported a rate of 16.6% rod

breakages in their series of 36 neuromuscular/syndromic patients treated with TGR extended to the pelvis. A rod breakage rate of 15 to 42% in TGR was published in early onset scoliosis from all etiology without pelvic fixation [32-34]. 10.6% were related to magnetically controlled growing rods [10]. In Mc Elroy series of growing rods for the treatment of scoliosis in cerebral palsy [4], mechanical complications occurred in 19/27 (70%) patients including rod exchange (6), rod fracture (5), anchor revision (4) or anchor dislodgement (2).

In the present series, despite the absence of PSF, rod breakage occurred in 5 patients at risk (cerebral palsy with dystonia or hyperactivity, ambulatory patients and high body weight). They were managed by simple rod replacement or the use of 4 rods construct in the lumbar area as shown in Figure 6.

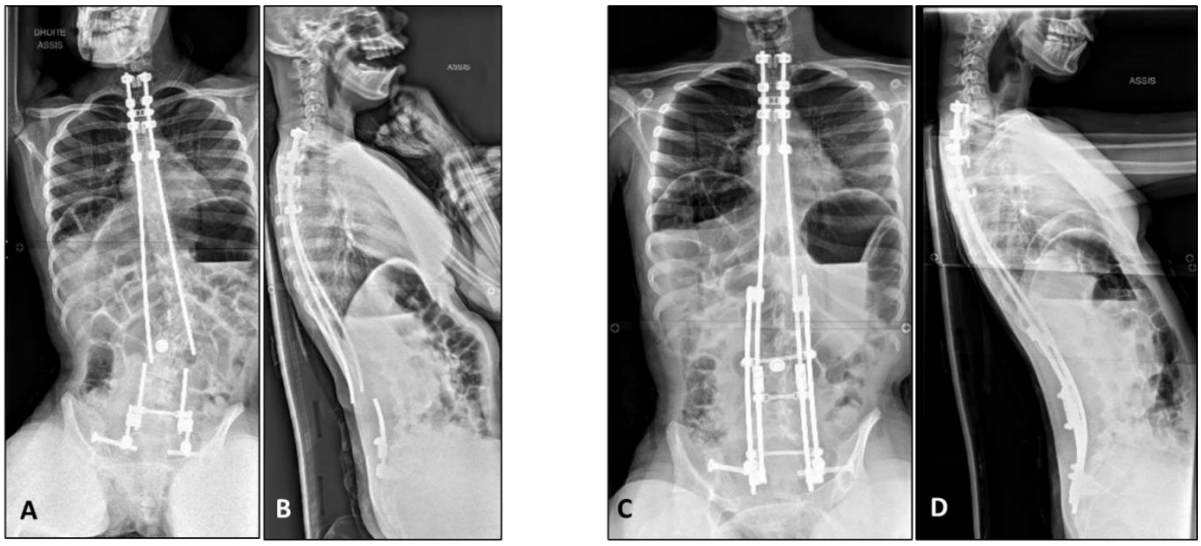

Figure 6 : Non-ambulatory 14 years old girl suffering from dystonic cerebral palsy . A/B : X-ray of rod fracture 4.5 years after initial surgery. C/D : X-ray after revision surgery and additional rod implantation.

For one of them, it was decided per-operatively not to replace the broken rod because of the observation of extensive auto fusion around the rod. The removal of the rod could have generated important damages. At 3 years after the rod breakage, the spinal deformity of this patient remained stable without any complaint. 3 cases of proximal hook dislodgement were

experienced, one due to a severe osteoporosis and the others because of a lack of rod contouring at the beginning of our learning curve. This low rate of proximal mechanical complications can be explained biomechanically by the stability of the double claw-hook construct [35]. We experienced 9 cases of proximal junctional kyphosis (11%) in our series without the need for revision surgery. This low incidence compared to other series [36] can be explained first by the type of the proximal fixation made of a solid double hooks claws; secondly by an adequate proximal bending of the rods; and finally by the high level of proximal limit of fixation (C7 or T1) in poor head control, sagittal trunk imbalance and hyperkyphotic cases.

The construct also draws its strength and stability from its pelvic fixation technique using ilio-sacral screws, which have proved their efficiency and reliability for more than 40 years of experience [29, 37]. No screw pullout or breakage were reported. Osteolysis around ilio-sacral screws was observed radiologically in 25 patients but no screw migration, pull-out or adverse clinical outcomes were associated. The screw loosening was due to the micro rotation of the screw concomitant to flexion/extension motion of the trunk of the patient. Ilio-sacral screw malposition was reported in 4 cases operated with freehand technique. No new case of misplaced screw was reported since the use of per-operative CT-scan control.

Finally, the infectious rate of 18% was lower than TGR treatment in patients with cerebral palsy (30%) [4] or spinal muscular atrophy [38], probably thanks to the minimally invasive approach preserving the intermediate area. The number of repeated surgical procedures is known as a high-risk factor especially in this population [39]. 3/15 infections occurred after initial surgery and 12 after rod lengthening procedures. This rate of infections after rod lengthening procedures could be reduced using self-expanding devices that avoid repeated surgeries.

One of the weaknesses of this study was the absence of comparative treatment to assess the lower rate of complications of the present surgical technique. Further work is under way to analyze long-term lung function and quality of life. Preliminary results suggest minimal impact

of surgery on pulmonary management, and an overall patient satisfaction. All patients still did not achieve their skeletal maturity and the study still needs long-term follow-up (>10years) to evaluate the effectiveness of this treatment to confirm the stability of the construct allowing the avoidance of PSF. Secondly, there were no radiological evidence to quantitatively assess the quality of the progressive ankylosis of the spine. These results open the way for further studies to evaluate spinal ankylosis using non-invasive exams (e.g, MRI and ultrasound).

CONCLUSION

The outcomes of the bipolar minimally invasive fusionless technique in this series with a mean of 6.5 years follow-up showed a stable and satisfying correction of spinal deformities and pelvic obliquity over time. The rigorous implantation of proximal and distal fixations contributed to reduce the rate of mechanical complications. In this series, half patients reached skeletal maturity and did not require PSF initially planned, leading to consider this technique as a viable alternative to PSF for neuromuscular scoliosis. However, longer follow-up is necessary to confirm these findings.

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A cinq ans de suivi post opératoire, nous avons pu observer une correction stable des déformations vertébrales et de l'obliquité pelvienne, avec un faible taux de complications mécaniques chez les 100 premiers patients opérés selon cette technique. Aucune arthrodèse vertébrale postérieure n'a été réalisée chez les patients de la série à maturité squelettique.

Néanmoins, il s'agit d'une population hétérogène (encéphalopathies, dystrophie musculaire, amyotrophie spinale infantile, et autres pathologies neuromusculaires non étiquetées), qui ne nous a pas permis de corréler les résultats au retentissement sur la fonction respiratoire et à la qualité de vie. C'est la raison pour laquelle, en complément de cette série historique des 100 premiers patients opérés, nous avons voulu étudier les résultats à long terme de l'instrumentation sans greffe bipolaire au sein d'une cohorte plus homogène de patients qui étaient tous atteints d'amyotrophie spinale infantile.

5.2. Article 2 : Minimally Invasive Fusionless Surgery for Scoliosis in Spinal Muscular Atrophy: Long-term Follow-up Results in a Series of 59 Patients.

Gaume M, Saudeau E, Gomez-Garcia de la Banda Marta, Azzi-Salameh V, Mbieleu B,Verollet D, Benezit A, Bergounioux J, Essid A, Doehring I, Dabaj I, Desguerre I, Barnerias C, Topouchian V, Glorion C, Quijano-Roy S, Miladi L.

J Pediatr Orthop. 2021 Oct 1 ;41(9):549-558.

Résumé

Contexte : Le traitement des scolioses chez les patients atteints d'amyotrophie spinale infantile (ASI) a évolué au cours de la dernière décennie, avec l'émergence des instrumentations sans greffe qui permettent de corriger la déformation avant la fin de la croissance. Ces techniques permettent de préserver la croissance du tronc en attente d'arthrodèse vertébrale postérieure à l'adolescence.

Objectif : Evaluer les résultats cliniques, radiologiques et respiratoires à long terme de la technique d'instrumentation sans greffe bipolaire chez les patients atteints d'ASI.

Patients et méthodes : Tous les enfants ASI atteints de scoliose et ayant été opérés selon la technique d'instrumentation sans greffe bipolaire dans notre service ont été inclus. Les données

cliniques, génétiques, respiratoires et radiographiques ont été revues rétrospectivement. Une enquête de satisfaction a été réalisée auprès des patients.

Résultats : 59 enfants avec confirmation génétique de ASI (9 ASI1c, 47 ASI2, 3 ASI3) ont subi une instrumentation sans greffe bipolaire avec un âge moyen de 11±1,9 ans lors de la chirurgie initiale. Aucun patient n'était marchant. 26 ont été traités par Nusinersen intrathécale. Le suivi moyen était de 5,2 ans. L'angle de Cobb moyen s'est améliorée de 79±15° à 41±16° et l'obliquité pelvienne a diminué de 24±11° à 5,9±4°. L'espace moyen disponible pour les poumons s'est amélioré de 77% à 93%. Des complications mécaniques ou infectieuses sont survenues chez 9 patients, avec retrait du matériel chez 1 patient. 6 enfants ont dû subir des ré interventions non programmées. Un corset postopératoire a été nécessaire chez 13 enfants. Le gain de poids moyen à 3 ans de recul était de 6 kg. 91,5 % des patients étaient satisfaits de la prise en charge. Il n'y a pas eu d'impact significatif sur la fonction respiratoire en post opératoire. Des retensions de tiges ont été réalisées chez 30 enfants, avec un intervalle moyen entre les procédures de 1,9 ans (0,5 à 3,7 ans). Aucun patient n'a eu besoin d'une arthrodèse au dernier recul.

Conclusion : L'instrumentation sans greffe bipolaire chez les patients ASI permet de préserver la croissance rachidienne et thoracique sans interférence avec la fonction respiratoire. Elle permet une correction significative de la déformation rachidienne et de l'obliquité pelvienne, avec un taux de complications réduit. La correction de la déformation de la colonne vertébrale a été maintenue à long terme, ne nécessitant pas de fusion définitive à la fin de la croissance.

Abstract

Background: Treatment of spinal muscular atrophy (SMA) scoliosis has evolved in the last decade, with the emergence of fusionless surgical techniques that allow correction of the deformity before the end of growth spurt. These techniques are expected to delay definitive spine fusion and preserve trunk growth.

Purpose: To evaluate long-term clinical, radiological and respiratory outcomes of a minimally invasive fusionless surgery (MIFLS) in SMA scoliosis.

Patients and Methods: Children affected with SMA scoliosis who underwent MIFLS in our department were included. The instrumentation consisted in a bilateral sliding rod construct from T1 to the sacrum, anchored proximally by double-hook claws and distally by ilio-sacral screws. Clinical, genetic, respiratory and radiographic data were retrospectively reviewed. A patient's satisfaction survey was performed.

Results: 59 children with genetic confirmation of SMA (9SMA1c, 47SMA2, 3SMA3) underwent MIFLS at a mean age of 11±1.9y. All of them were non-walker at the time of surgery. 26 were treated with intrathecal Nusinersen. Mean follow-up was 5.2y. Mean major coronal curve improved from 79±15° to 41±16° and pelvic obliquity decreased from 24±11° to 5.9±4°. Mean space available for lung improved from 77% to 93%. Mechanical or infectious complications occurred in 9 patients, with removal of the implant in 1. 6 children required unplanned surgeries. Postoperative bracing was needed in 13 children. Mean gain weight 3 years after the first surgery was 6kg. 91.5% of patients had a positive satisfaction of the surgery. There was no significant impact in respiratory function postoperatively. Only 30 children required rod lengthening procedures, with a mean interval between procedures of 1.9 years (0.5 to 3.7y). No arthrodesis was required at last follow-up in any patient.

Conclusion: Bipolar minimally invasive fusionless surgery in SMA preserves spinal and thoracic growth without interference with respiratory function. It provides a significant

correction of spinal deformity and pelvic obliquity, having a reduced rate of complications. The correction of spinal deformity was maintained at long-term, not requiring definitive fusion at the end of growth.

Level of evidence: 4

INTRODUCTION

Spinal muscular atrophy (SMA) is an autosomal recessive neuromuscular disorder caused by defects in SMN1 gene that lead to loss of anterior horn cells at the spinal cord. SMA is a progressive disease and shows variable clinical forms: SMA1 is the most severe form, in which infants do not achieve head or trunk support and most die in the first two years due to respiratory and bulbar muscle weakness [1]. Later onset is associated with less severe forms where patients acquire sitting (SMA2) or walking ability (SMA3). Those children who survive and do not achieve or lose walking ability will develop typically a pelvic asymmetry and a progressive spinal deformity which interferes with sitting position [2-4]. Scoliosis is also an important complication in these children because it may further impair respiratory function, which is already reduced due to intercostal muscle weakness and fatigue. The arrival of innovative and specific therapies in SMA is changing the prognosis and survival rate in particular in the most severe forms. Although improvement in motor function has been proven by different trials and in real-life studies, most patients do not achieve walking ability and scoliosis remains a major complication in them from very early ages.

Spinal fusion is generally preferred for the surgical treatment of SMA scoliosis, but fusion requires bone maturity and is not satisfactory before puberty. In young children, conservative treatment has been for decades the only treatment available of scoliosis. However, trunk and neck bracing is complex in SMA because the brace has to be rigid enough to maintain the trunk in a corrected position but not compressing the thorax, in order to preserve respiratory motion. Garches brace was conceived in France in the 60's to treat neuromuscular scoliosis [5] after the

polio epidemics. It is particularly adapted to treat SMA children from the age of one year [6]. While it is effective in the first years to treat axial collapse and prevent rapid impairment of the deformity [7,8], its benefits are less consistent at the time of growth spurt. Due to the severe deformities observed in SMA from early ages, many teams around the world have searched for fusionless surgical techniques in the last decades, with the aim of stabilizing the spine and at the same time preserving its growth, as well as of the thoracic cage [9-11]. So far, fusionless surgery studies in SMA have been limited to small sample sizes with short follow-up [12-13]. The latest and biggest series was published by Matsumoto et al. with an improvement of pulmonary function in 74 patients treated with growth-friendly instrumentation and 2 years follow-up [14]. A minimally invasive fusionless surgery (MIFLS) with a bipolar sliding construct was recently described in patients affected with neurological or neuromuscular diseases, including 22 SMA with 2 years follow-up [15]. Thanks to the less invasive approach compared with spinal fusion techniques and the high stability of the construct observed during follow-up, it rapidly replaced spinal fusion techniques in our department, in particular to treat neuromuscular scoliosis.

The purpose of this study was to evaluate long-term clinical, radiological and respiratory outcomes of MIFLS in a series of 59 children affected with SMA.

PATIENTS AND METHODS

Study design

The medical charts of 59 patients affected with SMA who underwent bipolar MIFLS for scoliosis, between June 2011 and November 2019 were retrospectively reviewed. All these patients were treated in our department which is the national reference center for neuromuscular diseases. Patients were classified into 3 groups [16]:

- SMA type 1, presented in the first few months of age with symmetrical limb weakness, intercostal muscle weakness, tongue fasciculations, and absent deep tendon reflexes.

- SMA type 2 presented between 6 and 18 months of age, with ability to sit but not to stand or walk dependently.

-SMA type 3, presented after 18 months and affecting children who achieve independent walking who but lose the ability over time.

The study was approved by the appropriate ethics committee (CPP, ID-EUDRACT #2014 A019043 44). Implants used were provided by Euros® Company, La Ciotat, France. The minimally invasive fusionless bipolar fixation technique was described in our department [15]. Two small midline skin incisions are made: one proximal at the upper thoracic spine and one distal at the lumbosacral junction. The fixation extends from T1 to the sacrum proximally anchored with hook claws [17] and distally by ilio-sacral screws [18,19]. The ilio-sacral screws are inserted percutaneously from the posterior part of the iliac bone to S1 body, in an oblique posterior-to-anterior direction avoiding the spinal canal. Pre-countoured medial long rods are inserted from proximal to distal incision in sub-fascial plan. They are fixed proximally in the hook claws and distally to lateral short rods by side-to-side closed connectors. The short rods are inserted and fixed inside the ilio-sacral connectors. The side-to-side connectors are first locked at the extremity of the medial long rods. The correction is obtained first by distraction maneuvers on the concave side between the ilio-sacral connector and the side-to-side connector that migrates proximally with the long rod. At the end of the distraction, the side-to-side connector is locked to the short lateral rod.

Next, similar rods are inserted and fixed in the convex side.

In case of incomplete correction of the pelvic obliquity after concave distraction, a compression maneuver is realized on the convex side before locking the side-to-side connector on the convex short rod.

This sliding construct allows the same corrective maneuvers during further rod lengthening procedures thought the distal incision by unlocking the side-to-side connectors of the short rods as shown on figure 1.

Rod lengthening procedures are proposed according to the following indications:

- to achieve further correction in case of residual spinal deformity or in case of curve worsening due to growth spurt.
- to improve pelvic balance in case of residual pelvic obliquity thanks to an asymmetrical rod lengthening allowed by the construct.
- in case of persistent trunk imbalance in the frontal or sagittal plane after initial surgery in severe cases.

The design of the construct and the absence of bone graft allow an easy intrathecal drug administration by radiologically guided lumbar puncture if needed.

Quality of life

A questionnaire was conceived to assess the difficulties faced by patients and caregivers during and after surgery, adapted from Comstock et al [20] and Bridwell et al [21]. Telephonic interviews were conducted by a clinician unknown to the families. Satisfaction with surgery was divided into two groups: positive (very satisfied and satisfied) and negative (neutral, unsatisfied and very unsatisfied). School level and presence of spinal pain were also asked.

Pulmonary function tests

Patient's pulmonary function was assessed by spirometry in supine position without the brace. Forced Vital Capacity (FVC) was performed, using a Vyntus™ SPIRO PC Spirometer. Difference between preoperative and latest FVC (in ml) have been reported as well as the interval of time between these two tests (in years).

The use of mechanic ventilatory support (nocturnal vs nocturnal and diurnal) and the type of ventilation (non-invasive vs invasive via tracheostomy) as well as the main events in the respiratory management such as intubation duration and number of days in the ICU (intensive care unit) ward were assessed.

Radiologic data

For each patient, the following radiologic parameters before surgery, immediately after surgery, and at last follow-up were reviewed: major coronal curve, pelvic obliquity, thoracic kyphosis, lumbar lordosis, coronal balance, T1-T12 height, T1-S1 height, space available for lung (SAL), chest width and chest depth. Radiologic measures were performed using previously described methods [22-24]. We also recorded any complication observed, including implant failure, infections and neurological complications.

Statistical analysis

Variables are described as mean values with a range (±SD). Changes in radiological values and pulmonary function tests were calculated by applying the pairwise Student's t test, from baseline to the early post-operative assessment and to last follow-up. P values below 0.05 were considered significant. Complications were described as n (%). The various analyses were performed using R (http://www.R-project.org) software.

RESULTS

Clinical data

Results are reported in Table 1.

59 consecutive children (29 females, 30 males) with genetic confirmation of SMA (9SMA1c, 47SMA2, 3SMA3) were included in the study. Mean follow-up was 5.2y (2y to 9.6y). They had a mean age of 11.0±1.9 years at surgery. SMN1 gene defects were confirmed in all the patients. 26 patients were treated in the course of the study by Nusinersen (SPINRAZA®). The injections were administrated intrathecally under radiological guidance (fluoroscopy or tomography). A minimal security period of four weeks was maintained in all injections before or after a surgical intervention (implant or rod lengthening). This period was prolonged in case of surgical complications. They were well tolerated and there were not secondary infections (meningitis, implant infection) or other complications (cerebrospinal fluid leaking) related with the lumbar punctures in any of the treated patients.

Six patients required blood transfusion after surgery.

Concerning motor function, none of the patients were ambulatory at the time of the study. Prior to surgery, 32 patients had a self-head control that was maintained post operatively. Most spinal deformities were "C" shaped thoracolumbar (27 right, 24 left) or thoracic (3 right, 1 left). They were occasionally "S" shaped (4). Mean preoperative body weight was 30.6Kg±10.9 and 35.5Kg±9.8 at latest follow-up. The main part of weight gain was observed in the 3 first years after surgery with a mean of 6.1Kg. Nine patients were fed via gastrostomy. No supplementary perioperative gastrostomy was performed in the period of the study. All patients followed a previous orthopedic treatment by a trunk orthosis, a Garches brace in most of them (54). Due to the severity and stiffness of the spinal deformity, ten patients required a cephalic halo traction preparation before surgery, with progressive increasing of traction weight during four to six weeks.

A total of 46 rod lengthening (RL) were performed in 30/59 patients (1 to 4 procedures per patient). Mean age at initial surgery was 10.9±1.7y. Mean interval between initial surgery and 1st RL was 1.9±0.8y. 2nd RL was performed after an interval of 1.5±0.5y years. Three patients required a 3rd RL after 1.5±0.1y, and one had a 4th RL.

Patients who did not require RL (29/59) had a mean age of 12.6±1.4y at initial surgery (11.7 to 15.1y) and a mean follow-up of 4.1±1.6y.

Quality of life

All patients attended school. 56% were in a normal level for their age, 8% in a school with rehabilitation facilities, 16% in secondary school, 12% in technical professional school and 4% studied at home. All patients were able to drive their electric wheelchair independently.

Satisfaction with surgery was positive (very satisfied or satisfied) in 54 patients (91.5%) and negative (indifferent, dissatisfied and very dissatisfied) in the remaining 5. Reasons for negative satisfaction were the maintain of bracing after surgery (1), the need of iterative rod lengthening (2), persistent lower back pain (1) and chronic infection requiring the removal of the instrumentation (1).

	Type 1c (n=9)	Type 2 (n=47)	Type 3 (n=3)	All types (n=59)
Age at initial surgery (years)	12.7±1.1	10.7±1.8	11.9±2.8	11.0±1.9
Pre-operative motor status				
Self-head control (n)	0	29	3	32
Ambulation (n)	0	0	0	0
Garchois brace (n)	9	42	4	55
Without chin strap (n)	0	4	0	4
Post-operative brace				
Garches brace (n)	5	6	0	11
Without chin strap (n)	1	1	0	2
Head support without brace	3	5	0	8
Total patients with brace at final	6/9	7/47	0/3	13/59
Type of curve (n)				
Left thoracic	0	1	0	1
Right thoracic	0	3	0	3
Left Thoraco lumbar	4	18	2	24
Right horaco lumbar	4	21	2	27
Double major	1	3	0	4
Body weight (kg)				
Preoperative	28.6±7.2	29.3±10.3	56.9±20.4	30.6±10.9
Last follow-up	36.4±5.6	34.5±9.5	60.7±17.3	35.9±10.0
Gastrostomy (n)	4	5	0	9
Halo traction device before surgery (n)	1	9	0	10
Total RL per patient (n)				
0	5	24	0	29
1	2	14	2	18
2	2	6	1	9
3	0	2	0	2
4	0	1	0	1
Interval between:				
Initial surgery -1st RL (years)	1.9±0.6	2.0±0.9	3.6±1.8	1.9±0.8
1st RL - 2nd RL	1.3±0.4	1.2±0.3	2.8	1.5±0.5
2nd RL -3rd RL	-	1.5±0.1	-	1.5±0.1
3rd RL-4th RL	-	0.3	-	0.3

Table 1: Main clinical data of the series

RL : rod lengthening

Back pain was not reported during long-term follow-up in 52 patients. In the remaining seven, back pain was relieved by analgesics (3), was only mechanical, during mobilisation from toilets to wheelchair or to bed (3) or appeared only at the end of the day (1).

The questionnaire is included as Supplement 1.

Radiologic data

Table 2 and 3 details all the changes in radiological measures before, after surgery and at last follow-up.

Mean major coronal curve improved from 79±15° preoperatively to 39±14 postoperatively and 41±16° at last follow-up. Pelvic obliquity decreased from 24±11° preoperatively to 5.8±4 postoperatively and 5.9±4° at last follow-up.

Mean T1-T12 height increased from 20.1±2.7 to 23.6±2.5cm immediately postoperatively and reached 24.9±2.4cm at last follow-up. Mean T1-S1 height increased from 29.6±3.5 to 34.6±3.5cm immediately postoperatively and reached 36.5±3.1cm at last follow-up.

	Type 1c (n=9)	Type 2 (n=47)	Type 3 (n=3)	All types (n=59)
Major coronal curve (degree)				
Pre-operative	70.6±16.0	82.6±15.0	63.4±11.3	79.8±15.8
Post-operative	39.8±10.0	39.1±14.3	39.6±14.3	39.2±14.1
Last follow up	40.9±14.2	41.8±16.2	41.4±16.2	41.7±16.1
Thoracic kyphosis (degree)				
Pre-operative	34.8±12.8	43.3±18.8	26.0±6.2	41.0±18.0
Post-operative	22.2±8.5	27.0±8.1	28.3±10.5	26.4±8.5
Last follow up	21.8±8.7	25.4±6.7	27.2±12.7	25.0±7.5
Kyphosis > 50° (n)				
Pre/ Post / Last follow up	2/0/0	15/2/0	0/0/0	17/2/0
Lumbar lordosis (degree)				
Pre-operative	27.9±7.5	36.8±11.3	51.8±12.5	36.4±12.0
Post-operative	28.9±7.5	35.6±8.2	48.2±2.1	35.3±8.7
Last follow up	33.5±4.0	38.5±7.9	38.1±1.1	37.8±7.0
Coronal imbalance (mm)				
Pre-operative	50.5±3.1	26.8±2.3	29.5±0.7	30.1±2.4
Post-operative	15.5±0.7	17.7±1.5	0	16.7±1.4
Last follow up	10.9±1.3	11.4±1.3	0	10.9±1.3
Pelvic obliquity (degree)				
Pre-operative	23.4±5.6	24.2±12.5	24.6±11.5	24,1±11.4
Post-operative	4.6±3.5	5.9±4.6	6.7±6.3	5.8±4.4
Last follow up	5.1±2.5	6.0±4.4	6.4±5.8	5.9±4.3
Pelvic obliquity > 15° (n)				
Pre/ Post / Last follow up	7/0/0	25/1/0	2/0/0	34/1/0
T1-T12 height (cm)				
Pre-operative	22,1±2.4	19.5±2.5	23.3±1.5	20.1±2.7
Post-operative	25.3±2.8	23.2±2.3	24.4±2.4	23.6±2.5
Last follow up	26.5±2.0	24.6±2.2	25.8±3.1	24.9±2.4
T1-S1 height (cm)				
Pre-operative	32,2±4.1	29.1± 3.2	30.5±3.3	29.6±3.5
Post-operative	38.0±2.7	34.0±3.4	34.1±2.6	34.6±3.5
Last follow up	38.8±2.4	36.2±3.5	35.8±1.55	36.5±3.1
SAL ratio				
Pre-operative	0.80±0.06	0.75±0.10	0.92±0.14	0.77±0.1
Post-operative	0.91±0.07	0.90.3±0.06	0.93±0.04	0.90±0.06
Last follow up	0.91±0.05	0.93±0.05	0.98±0.01	0.93±0.05
Chest width (mm)				
Pre-operative	136.7±35.5	133.7±24.6	136.3±13.5	134.3±25.9
Post-operative	147.4±33.3	149.1±19.3	161.6±41.5	149,6±22.4
Last follow up	168.5±20.5	167.1±2.2	173.0±1.3	163.9±24.5
Chest depth (mm)				
Pre-operative	107.2±19.3	114.5±28.3	127.0±2.0	113.9±26.4
Post-operative	139.0±16.5	123.9±28.3	148.0	126.6±19.7
Last follow up	134.7±8.7	132.9±23.8	173.5±1.5	135.3±23.0

Table 2: Mean radiographic data of the series.

SAL: Space available for lung; RL : rod lengthening

	Without RL (n=29)	≥1 RL procédure (n=30)	*p-value*
Age at initial surgery (years)	12.6±1.4y	10.9±1.7y	<0.05
Major coronal curve (°)			
Preoperative	79.5±16.1	79.9±16.4	>0.05
Postoperative	38.1±14.6	38.4±14.5	>0.05
Last follow-up	39.1±16.9	41.5±16.9	>0.05
Pelvic obliquity (°)			
Preoperative	23.4±11.1	23.7±11.3	>0.05
Postoperative	5.5±4.8	5.8±4.5	>0.05
Last follow-up	6.1±4.3	6.3±4.3	>0.05

Table 3: Mean major coronal curve and pelvic obliquity according to the number of rod lengthening procedures and age at initial surgery

RL : rod lenghtening

Complications

Table 4 shows the type and characteristics of complications in the series. No patient had a fatal complication in the perioperative period or during follow-up. Nine children presented mechanical and/or infectious complications (8 and 5 patients respectively). Six children required unplanned surgeries. Infections were successfully treated by irrigation, debridement, and appropriate antibiotic therapy in four patients. One child required implant removal because of chronic infection.

	Initial surgery (n=59)	1st RL (n=30)	2nd RL (n=12)	3rd RL (n=3)	4th RL (n=1)	Total
Blood transfusion	6	0	0	0	0	6
Intraoperative cardiac arrest (due to PNO, full recovery)	1	0				1
Wound infection	3	0	1	0	1	5
Mechanical complication						
Proximal hook dislodgment	1	0	0	0	1	2
Proximal rod prominence	0	0	1	0	0	1
Prominence of IS screw	1	0	0	0	0	1
IS malposition	0	1	2	1	0	4
Neurologic deficit	0	0	0	0	0	0
Meninges breach	0	1	0	0	0	1
Unplanned surgeries (n)	5	0	1	0	0	6

Table 4: Complications

RL = rod lengthening; PNO: Pneumothorax; IS: Ilio-Sacral

Pulmonary function data

Pulmonary function data are reported in Table 5.

	Type 1c (n=9)	Type 2 (n=47)	Type 3 (n=3)	All types (n=59)
Preoperative Ventilatory support				
Tracheotomy (n)	6	1	0	7
NVI	2	19	0	21
Additional supports	0	29	0	29
Immediate extubating (n)	2/9	23/47	3/3	28/59
Intubation ≥1 day, (n, mean)	1/9 (7 days)	23/47 (5.5±3.2)	0	24/59 (5.5±2.4)
Post-operative Ventilatory support				
Tracheostomy (n)	6/9	1/47	0	7/59
NVI	2	19	0	21
ICU stay required (days)				
Initial surgery (n=59/59)	14.8±6.1(n=9)	11.7±7.1(n=46)	11.0±8.5(n=4)	11.8±7.6
1st RL (n= 19/30)	10.25±7.8(n=4)	7.3±5.6(n=14)	3.0 (n=1)	7.7±6.1
2nd RL (n=4/12)	3.0 (n=1)	3.3±1.5 (n=3)	-	3.2±1.2
3rd RL (n=3/3)	-	4.5±1.5 (n=3)	-	4.5±1.5
4th RL (n=1)	-	4.0 (n=1)	-	4.0
	Type 1c (n=7)	Type 2 (n=39)	Type 3 (n=3)	All types (n=49)
Mean pre-operative FVC (ml)	804±467	1015±571	2223±240	1059±620
Mean post-operative FVC (ml)	790±389 (p<0.05)	941±566 (p<0.05)	2563±368 (p<0.05)	1019±633 (p<0.05)
Mean interval between FVC (years)	2.1±1.4	3.9±2.1	2.9±1.2	3.5±2

Table 5: Respiratory outcomes

RL = rod lengthening

7 patients were ventilated via tracheostomy preoperatively. No patient required a peri- operative tracheostomy or in the following years. Concerning the initial surgery, 28 patients were extubated the same day of the procedure. For the other patients, mean intubation duration was 5.5±2.4 days, with a mean stay in the ICU of 11.8±7.6 days. After the first RL, 19/30 patients were managed in the ICU, with a mean stay of 7.7±6.1 days. Mean duration of hospital stay at

the orthopedic ward was 5.5±4.0 days. Only 4/12 patients requiring a second RL were admitted in the ICU and they stayed for a mean of 3.2±1.2 days followed by a mean duration of hospital stay at the orthopedic ward of 5.4±1.6 days.

49 of the 59 (83 %) patients included had reliable pre and postoperative FVC testing (7 SMA1c, 39 SMA2 and 3 SMA3).

DISCUSSION

Spinal deformities in SMA occurs in the childhood and need early treatment to prevent severe complications [25-26]. When conservative methods (external bracing, spinal casting, cephalic halo traction) are not efficient to prevent curve progression, surgical treatment is required. Until recently, spinal fusion was the most recommended procedure to limit severe deformity progression but was not a satisfying option in early ages because of arrest of spinal and thoracic growth with pejorative consequences [27]. To allow surgical management before the end of skeletal maturity, several fusionless techniques (e.g., traditional growing rods (TGR) or magnetically controlled growing rods (MGCR), vertical expandable prosthetic titanium rib (VEPTR) have been developed in the last decades, but a high rate of complications is reported in SMA population [28-31]. In a recent series published by Swarup et al. [32], the implant related complication rate reached 23% with MGCR and up to 47% with VEPTR. These techniques are provisory and usually followed by arthrodesis at older age.

To our knowledge, the present study is the largest homogeneous monocentric series, with the longest follow-up of SMA scoliosis treated by the same technique. MIFLS bipolar technique differs from TGR as it is definitive thanks to a solid construct relying on strong proximal and distal fixations. The stability of the results at long-term and the absence of rod breakage made it possible to avoid the final arthrodesis for all patients, that was initially planned [33]. This technique became the standard of surgical treatment for neuromuscular scoliosis in our

department. However, even if this construct is definitive, it remains evolutive as it allows a gradual correction of the spinal deformity thanks to further symmetric or asymmetric rod lengthening procedures, if necessary, to improve the correction of residual spinal deformity or pelvic obliquity.

In opposition to TGR, the auto-fusion occurs at a delayed stage in the bipolar construct thanks to the preservation of the intermediate area between the two fixations. Thus, the avoidance of early fibrosis allowed a mean interval of rod lengthening procedures of 1.6 years in our series instead of 6 months as recommended for TGR [34]. However, 29/59 patients who were operated at an older age (>12 years) did not require lengthening of the rods with satisfying and stable correction of the spinal deformity.

The mean age of 11 years at initial surgery was older than other series and was related to the systematic use in our multidisciplinary team of Garches brace, a rigid trunk orthosis especially designed for neuromuscular patients with respiratory insufficiency [6]. The spinal stiffening may be the achievement of a progressive and extended auto-fusion induced by the metallic rods implanted for many years. This hypothesis is already known and was supported by Jain et al. [35] who reported that avoiding final surgical fusion at skeletal maturity was a viable option for patients treated with growing rods with satisfactory final alignment and trunk height. Rewais et al. [12] suggested that definitive fusion in non-ambulatory SMA patients may be unnecessary after posterior distraction by growing rods. In Mc Elroy series, 40% SMA required an arthrodesis with a mean follow up of 4.5y [36].

The avoidance of arthrodesis should contribute to the benefits of spinal surgery on pulmonary function through adulthood. Long term follow-up studies of posterior fusion in SMA have suggested a continued deterioration of pulmonary function postoperatively [37,38] even if children treated with prior growth-friendly spinal implants have a better correction of their spine

deformity after spinal fusion in comparison to previously untreated patients [39]. Only one study has demonstrated significant improvement of respiratory function at final follow-up in SMA patients treated with surgical spinal stabilization by Harrington instrumentation or segmental spinal instrumentation [40].

In the present series, there was no significant impact in the respiratory function and patients continued their usual pulmonary management in accordance with the national standards of care of SMA [41], consisting of ventilation when indicated and in-exsufflator therapy once or twice daily for a minimum of 20 minutes [42]. Average chest width, space available for lung and trunk height increased over time. All of these radiographic findings are in accord with the overall favorable pulmonary results. In a series of 15 SMA patients treated with traditional growing rods, McElroy and al. [36] reported an increased rib angle but they did not measure the vital capacity to evaluate the presence of a restrictive syndrome. Chandran et al. [43] reviewed 11 patients with SMA for a mean follow-up of 9 months. Only 4 of 11 patients had available pre and postoperative FVC values, with an average of 0.53L preoperatively, 0.67L postoperatively and an increased Forced Expiratory Volume.

In the last years, innovative treatments have emerged in SMA, and the almost invariable mortality observed in the pre-therapeutical era for the most severe forms (SMA1c) has evolved to a higher survival of patients [44]. In the present cohort, almost half of the patients were treated by one of the new therapies, Nusinersen, an oligonucleotide intrathecally administrated. They were all treated at a late stage of the disease. The MIFLS bipolar technique allowed an easy intrathecal injection of the drugs thanks to the absence of posterior fusion and the free access to lumbar intrathecal space through the construct. They were guided by fluoroscopy or tomography and no infectious, neurological, or traumatic complications were observed. A security period of a month before or after the surgical procedure was respected.

Very remarkably there was a large reduction of the severity of the curve after surgery which was maintained at long-term. Kyphosis was also found to be effectively controlled in all patients. Thanks to the asymmetric rod lengthening allowed by the sliding construct and the pelvic fixation with ilio-sacral screws, all patients had a well-balanced pelvic obliquity and thus a more comfortable sitting position (Figure 1 and 2).

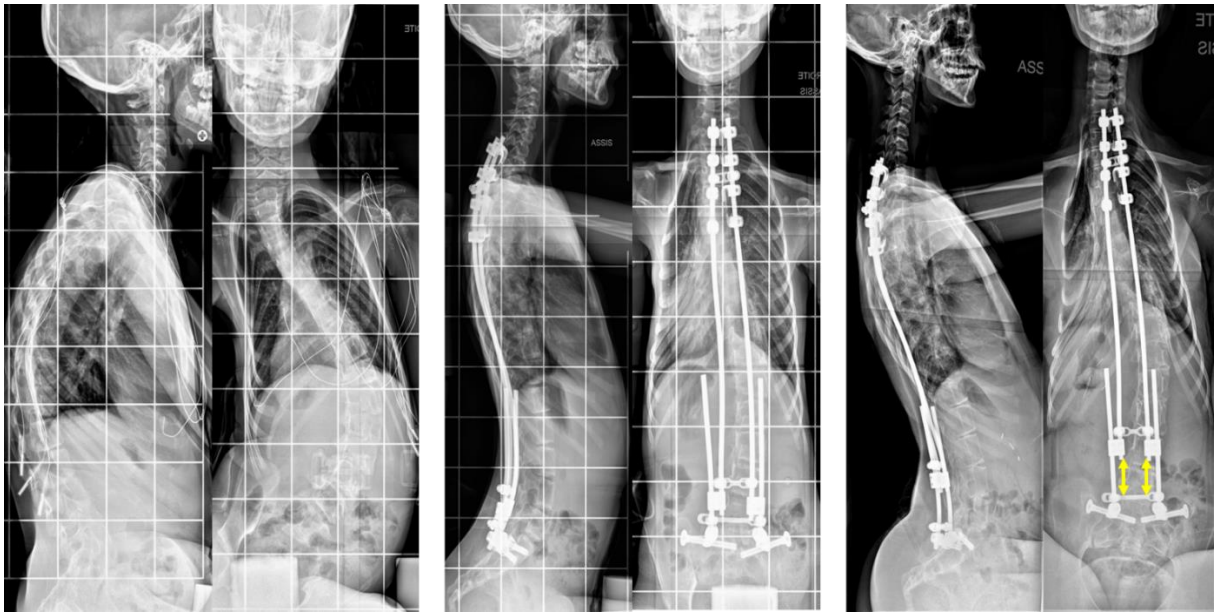

Figure 1: Radiographs of a girl with spinal muscular atrophy A/B, Preoperative x-rays C/D X-rays after initial surgery. E/F, X-rays after 4years follow-up and one rod lengthening procedure (yellow arrows)

Figure 2: A/B Pre- and C/D postoperative photographs of this patient.

The quality of life was improved, and positive satisfaction was reported by more than 90% of parents and patients. Continuation of brace use was not necessary for most of them. Autonomy, functional general status were significantly improved and a marked gain of weight in the 3 years following surgery was observed. Negative satisfaction of the surgery was only reported in 5 cases and was mainly related with the occurrence of infectious complications or with the impossibility of avoiding bracing postoperatively. However, this last reason was expected in SMA1c or severe SMA2 children because of pre-existing cervical weakness and inability for the head control before surgery. These results stress the importance of dealing with expectations of patients and their families preoperatively, concerning the potential complications following surgical procedures and the need of maintaining neck orthesis in the more severe patients.

Despite the efficiency and the satisfactory results of the bipolar MIFLS technique used in this series, the main drawback remains the need of repeated surgical procedures for rod lengthening in young patients. To avoid such surgical rod lengthening, MCGR are increasingly used but the complication rate is still high at long follow-up. Also, the difficulties of MCGR contouring on

the sagittal plane due to the presence of the internal magnetically drivable lengthening mechanism make this device less adaptable to the lordotic curve that are common in SMA scoliosis. MGCR were proposed by Lorenz et al. [45] in 21 SMA2 but 6 lengthening procedures failed, with 3 patients requiring surgical revision. In Colombo et al. MCGR series, stable pulmonary function was maintained but only 4 SMA2 were included [46].

To avoid iterative lengthening procedures with the bipolar MIFLS technique, a self-expanding rod was recently associated to the construct and has been evaluated in a clinical study of 20 patients (ClinicalTrial.gov: NCT02266667), including 4 SMA cases, with encouraging results [47].

Since arthrodesis was not required in any patients treated by the bipolar MIFLS technique in the present series with a long follow-up of almost 10 years for some of them, the advent of the self-expanding rod avoiding repeated surgeries could make the "bipolar MIFLS self-expanding technique" a treatment particularly adapted to the SMA population that we could propose now at an earlier age (<10 years old) than in the present series (11.0±1.9).

CONCLUSION

The bipolar minimally invasive fusionless surgery used in this series of 59 SMA scoliosis provided significant and stable correction of spinal deformities and pelvic obliquity, while preserving respiratory function, spinal and thoracic growth. Quality of life, general and functional status of the patients were improved. The avoidance of final fusion at long follow-up allowed by this technique could make it a viable alternative to arthrodesis in SMA population.

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Appendix 1 : Telephonic interview

Minimally Invasive Fusionless Surgery for Scoliosis in Spinal Muscular Atrophy: Long-term Follow-up Results in a Series of 59 Patients.

Telephonic interview

Patient Label

Date of evaluation:
Date of birth:
Date of initial surgery:

Male/Female
Etiology: SMA1 SMA2 SM3
Body weight: Height: Gastrostomy Y/N Tracheotomy Y/N

1. Motor status

Self-head control	Yes	No
Ability to sit	Yes	No
Ambulation	Yes	No
Ability to drive an electric wheelchair	Yes	No

2. Satisfaction with surgery

Raison for negative satisfaction:

Need of brace after surgery	
Repeated surgery for:	
- repeated rod lengthening	
- mechanical or infectious complication	
Persistent lower back pain	

3. Presence of back pain

Yes / No
-Relieved by analgesics: Y/N
-Circumstances:

4. Education

Enrolled in a school	
Normal level for the age	
School with rehabilitation facilities	
Secondary school	
Technical professional school	
School at home	

La technique d’instrumentation sans greffe a permis de préserver la croissance rachidienne et thoracique, sans interférence avec la fonction respiratoire. Le ressenti de la chirurgie a été positif, avec un maintien de la scolarité et peu de plaintes douloureuses. Elle a permis une correction significative de la déformation vertébrale et de l'obliquité pelvienne, avec également un taux réduit de complications. La correction de la déformation vertébrale a été maintenue à long terme, et aucune arthrodèse vertébrale postérieure n’a été proposée à la fin de la croissance.

Si ces deux cohortes renforcent notre hypothèse de la survenue d’un enraidissement progressif du rachis au cours du temps chez ces enfants, qui pourrait éviter une arthrodèse vertébrale postérieure à maturité squelettique, il est essentiel de faire le lien entre ces résultats cliniques et la fusion osseuse obtenue à la fin du programme de croissance.

Dans le chapitre suivant, nous avons évalué par tomodensitométrie, les modifications osseuses après instrumentation sans greffe bipolaire, à plus de cinq ans de recul.

S’agissant d’une population pédiatrique, en accord avec nos convictions éthiques, nous n’avons pas réalisé de TDM dans un but de recherche. Nous avons cependant pu recueillir 10 TDM réalisés à plus de cinq ans de recul pour d’autres motifs médicaux. L’article a été accepté dans la revue *European Spine Journal* (Avril 2023).

6. Analyse osseuse scanographique

6.1. Article 3 : Spontaneous induced bone fusion in minimally invasive fusionless bipolar fixation for neuromuscular scoliosis: A computed tomography analysis

M Gaume, T Langlais, K Loiselet, S Pannier, W Skalli, C Vergari, L Miladi

Résumé

Objectif : L'arthrodèse vertébrale postérieure (AVP) à la maturité du squelette reste la technique de référence chez les enfants atteints de scoliose neuromusculaire (SNM) après instrumentation sans greffe. Le but de cette étude par tomodensitométrie (TDM) était de rechercher et de quantifier une fusion osseuse spontanée à la fin d'un programme d'allongement par instrumentation sans greffe bipolaire.

Méthodes : Les SNM opérés par instrumentation sans greffe bipolaire de T1 au bassin en fin de programme de retension ont été inclus. Un TDM a été réalisé au moins cinq ans après l'opération. L'autofusion était rapportée comme complète ou non au niveau des facettes (dans les plans coronal et sagittal, à droite et à gauche, de T1 à L5), et autour des tiges (dans le plan axial, à droite et à gauche, de T5 à L5). Les hauteurs des corps vertébraux ont été évaluées.

Résultats : Dix patients ont été inclus (10,7 ans, sd2 lors de la chirurgie initiale). L'angle de Cobb moyen était de 82,5°, sd20 en préopératoire et de 37,6°, sd13 au dernier suivi. Le TDM a été réalisé en moyenne 6,7 sd 1,7ans après la chirurgie initiale. La hauteur moyenne préopératoire et lors du dernier recul des vertèbres thoraciques était respectivement de 13,5mm, sd1,7 et de 17,4mm, sd1,7 ($p<0,001$). 93% des articulations postérieures étaient fusionnées (sur 320 articulations analysées), correspondant à 15/16 niveaux vertébraux. L'ossification autour des tiges a été observée dans 6,5, sd2,4 niveaux sur 13 du côté convexe, et 4,2,sd2,2 du côté concave ($p=0,04$).

Conclusions : Il s'agit à notre connaissance de la première étude TDM quantitative qui a permis de mettre en évidence une autofusion spontanée après instrumentation sans greffe bipolaire, tout en préservant la croissance vertébrale. Cela pourrait être un argument supplémentaire pour remettre en question la nécessité d'une AVP à la maturité du squelette.

Abstract

Purpose: Posterior spinal fusion (PSF) at skeletal maturity is still the gold standard in children with neuromuscular scoliosis (NMS) who underwent fusionless surgery. The aim of this computed tomography (CT) study was to quantify the spontaneous bone fusion at the end of a lengthening program by minimally invasive fusionless bipolar fixation (MIFBF), that could avoid PSF.

Methods: NMS operated on with MIFBF from T1 to the pelvis and at final lengthening program were included. CT was performed at least five years postoperatively. The autofusion was classified as completely or not fused at the facets joint (on both coronal and sagittal plane, right and left side, from T1 to L5), and around the rods (axial plane, right and left side, from T5 to L5). Vertebral body heights were assessed.

Results: Ten patients were included (10.7y,sd2 at initial surgery). Mean Cobb angle was 82.5°,sd20 preoperatively and 37.6°,sd13 at last follow-up. CT were performed on average 6.7y,sd1.7 after initial surgery. Mean preoperative and last follow-up thoracic vertebrae height

were respectively 13.5mm, sd1.7 and 17.4mm, sd1.7 ($p<0.001$). 93% facets joints were fused (out of 320 analyzed joints), corresponding to 15/16 vertebral levels. Ossification around the rods was observed in 6.5,sd2.4 levels out of 13 in the convex side, and 4.2,sd2.2 in the concave side ($p=0.04$).

Conclusions: This first computed quantitative study showed MIFBF in NMS preserved spinal growth, while it induced 93% of facet joints fusion. This could be an additional argument when questionning the real need for PSF at skeletal maturity.

INTRODUCTION

Neuromuscular spinal deformities are related to abnormality in muscle tone and trunk imbalance, which can cause a loss of sitting ability. They often start in the first years of life [1, 2]. When conservative treatment is no more effective, various surgical strategies are proposed for the treatment of children with neuromuscular scoliosis (NMS) [3].

Early posterior spinal fusion (PSF) can cause growth inhibition and has negative effect on pulmonary function in young children, with decreased vital capacity and restricted pulmonary disease [4, 5]. Moreover, it does not arrest progression of the spine deformity even after anterior and posterior fusion [6, 7]. Difficulty in curve control after early fusion may be explained by crankshaft phenomenon defined as worsening of rotational deformity in young patients with solid posterior fusions resulting from persistent anterior growth of the vertebral bodies [8]. Zhang et al. also suggested the role of neurocentral synchondroses that creates a scoliotic deformity in growing-animal model correlated with the degree of closure of the synchondrosis [9].

Another treatment option is fusionless surgery which is considered more physiologic as it preserves the growth and motion of the spine and chest. Innovative fusionless techniques have been developed over the past decades (traditional and magnetically controlled growing rods, vertebral body tethers …) with satisfactory results [10, 11]. However, the management at the end of the lengthening program is still controversial. Balsano et al. [12] recommend PSF at skeletal maturity even in case of good and satisfying final correction. The complication rate

after such final fusion following traditional growing rods program is nevertheless high, reaching 20% in Poe-Kochert et al series [13]. Moreover, PSF has a low effect on spinal deformity improvement, and usually achieved <50% additional correction of the deformity remaining at the end of the growing-rod management [14]. Risk factors for reoperation following final fusion after GR program have been studied by Du et al. [15]. On the contrary, Jain et al. [16] avoided PSF in patients who have satisfactory final alignment and trunk height, a minimal gain in length at the last distraction, and no clinical or radiographic evidence of implant-related problems. Bouthors et al. [17] considered retaining dual GR instead of PSF in patients at skeletal maturity with satisfactory deformity correction.

Recently, Gaume et al. [18] reported their results and complications of 100 neuromuscular patients with a mean follow-up of 6.5 ± 0.7 years after minimally invasive fusionless bipolar fixation. PSF was not performed for any patient due to the stability of the construct and of the results over time and the hypothesis of a progressive ankylosis of the spine.

However, there are presently no objective imaging data supporting the quantity of auto fusion of the spine to assess the avoidance of PSF decision. The aim of this study was to evaluate growth and bone fusion on computed tomography at the final lengthening program.

PATIENTS AND METHODS

Patients

A total of 10 neuromuscular children who underwent a typical MIFBF and reached the end of their lengthening program were included. They all had a preoperative thoracic CT-scan, and a thoraco-lumbo-pelvic computed tomography scans (TAP-CT) that was indicated for other reasons excluding spinal disorder. The TAP-CT was performed after a minimum of 5 years follow-up after MIFBF. TAP-CT-scan was not performed on routine for the follow-up of NMS patients because of the risk of irradiation [19].

Operative technique

The minimally invasive FBF technique was described in a previous paper [20]. Briefly, this construct has a proximal foundation with two pairs of hook claws [21] and caudally a foundation with a rigid fixation with two ilio-sacral screws [22-24]. The longitudinal members consist of four Titanium rods overlapping caudally where the rods are coupled with connectors and here the repetitive distractions are performed. The study was approved by the appropriate ethics committee (CPP, ID-EUDRACT #2014 A019043 44). Informed consent was obtained from parents/guardians of all participants.

Methods of assessment

Collected clinical data included age at first surgery, sex, diagnosis, number of lengthening procedures, duration of follow-up, and complications. Patients underwent helical TAP-CT with 1.25 mm slice thickness. Facet joints between T1 and L5 were classified as completely or not fused by analyzing them on coronal and sagittal plane at all vertebral levels. A fusion score was defined as the percentage of number of fused facet joint/ number of facet joints x100.

The rod induced ossification was assessed on axial plane at each spinal level below the proximal fixation, from T5 to L5, on the concave and convex sides.

Thoracic and lumbar vertebral height, from T1 to L5, as well as discal height, before and at latest follow-up were calculated as the mean of maximal anterior, posterior, left and right height in sagittal and coronal plane [25]. These measurements were compared to values obtained from the literature for healthy subjects [26].

All measurements were repeated by two experienced orthopedic surgeons using Carestream Vue PACS software (Carestream Health). Blind comparison between the two observers was performed to assess the reliability of the method. Discrepancies were resolved by consensus.

Additional analyses were reviewed before surgery and at last follow-up: major curve angle according to the Cobb method on the coronal plane [27], pelvic obliquity, thoracic kyphosis (T4-T12) and lumbar lordosis (L1-S1) on sagittal plane.

Statistical analysis

Continuous variables were presented as mean (Standard Deviation). The normality was verified with the Shapiro-Wilk test and differences were analyzed by the student's paired t-test or Wilcoxon signed-rank test if the normality condition was not verified. Alpha risk was set to 5% ($\alpha = 0.05$). Statistical analysis was performed with EasyMedStat (version 3.19; www.easymedstat.com).

RESULTS

Population

10 patients (six boys and four girls; four cerebral palsy, two spinal muscular atrophy type 2 and four syndromic) were included with a mean age at surgery of 10.7 years (y) (SD = 2y; ranging from 6 to 14.5 y). All patients were Risser 0 at initial surgery .The average follow-up was 9.2y (SD = 1.1; ranging from 8 to 11y). Seven thoraco-lumbar scoliosis, four double curve and two thoracic scoliosis were recorded with a mean preoperative Cobb angle of 82.5±19° (ranging from 50 to 118°). Mean pelvic obliquity was 20±11° preoperatively and 2±2.4°at latest follow-up. The pre-operative and last follow-up radiographic parameters are reported in table 1. The number of rod lengthening was on average three per patient (ranging from 1 to 4) with an average interval between procedures of 1.9±0.8 y. The global complication rate was 23% (3 patients). One upper hook pullout, one hook migration and one deep infection were reported.

Parameters	Preoperative	Last follow-up	p-value
Cobb angle (°)	82 (SD=20)	37 (SD=12)	<0.001
Thoracic kyphosis (°)	48.4(SD=27.2)	32.6 (SD=6.8)	0.09
Lumbar lordosis (°)	40.2 (SD=18.3)	37.4 (SD=10.1)	0.6
Pelvic obliquity (°)	19 (SD=17.5)	0.5 (SD=2.7)	0.002

Table 1: Pre and postoperative radiographic parameters

Computed tomography outcomes.

Computed tomography were performed on average 6.7 years (SD = 1.7y; ranging from 4.5 to 10y) after the first surgical procedure. Patients were 17.4±2y when the CT scan was performed, and all reached skeletal maturity (Risser 5).

The number of levels of facet joints fused was similar on right and left side (p=0.7), averaging 15 levels out of 16 (94% of assessed facet joints, ranging 84.3 to 100%).

Mean number of levels with ossification induced by the rod on the convex side and concave side were respectively 6.5 (SD = 2.4) and 4.2 (SD = 2.2) with a mean difference of 2.3 (SD = 3.1 CI95% = 0.2; 4.4; p = 0.04). Ossification around the rods was qualitatively confirmed by CT scan (Figure 1) and 3D reconstruction of the bone by CT-scan thresholding (Figure 2-3).

Figure 1A/Sagittal CT scan view with rod induced ossification B/Sagittal CT scan view with lumbar joint fusion.

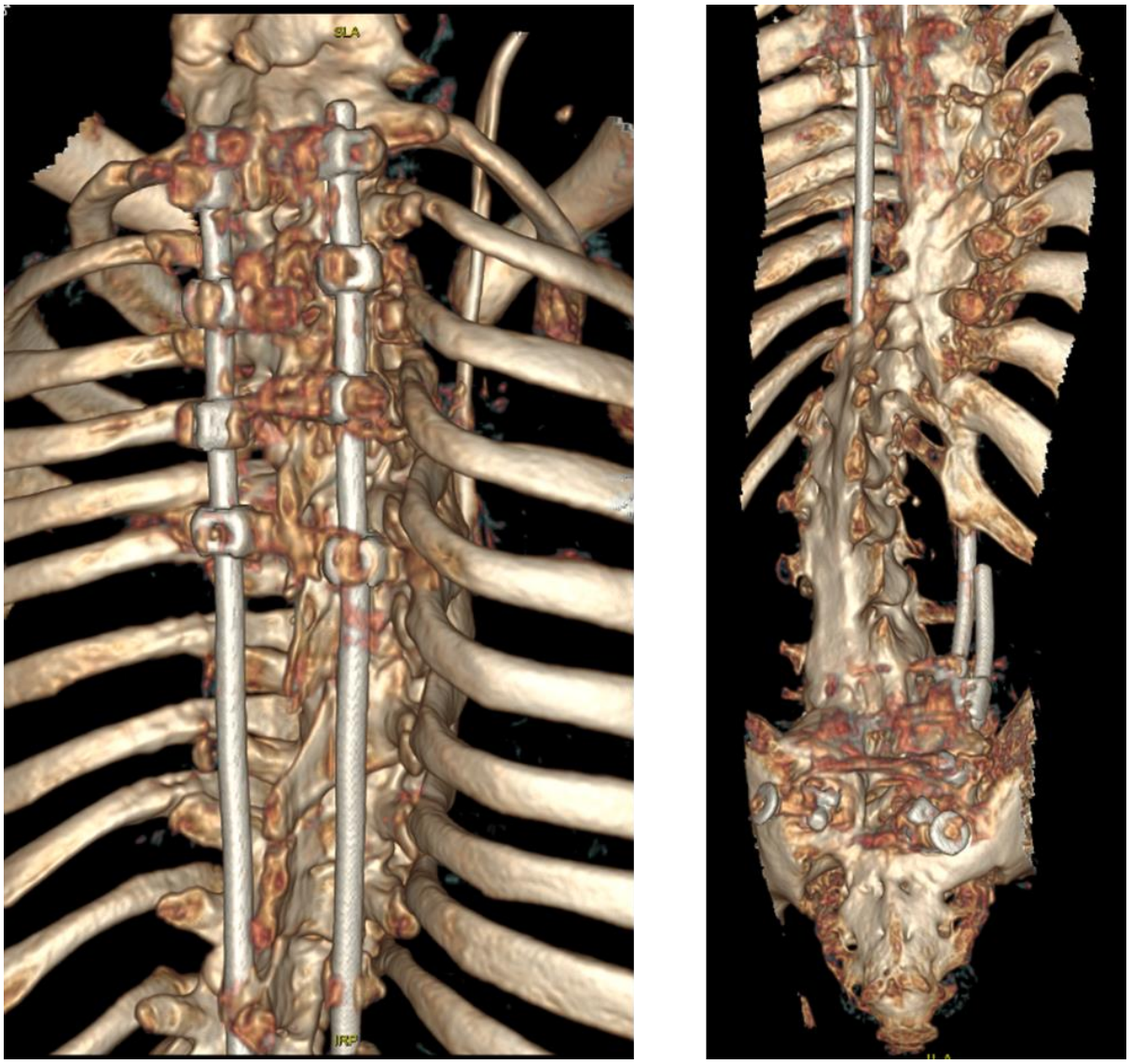

Figure 2 : 3D reconstruction with rod induced ossification and articular joint fusion.

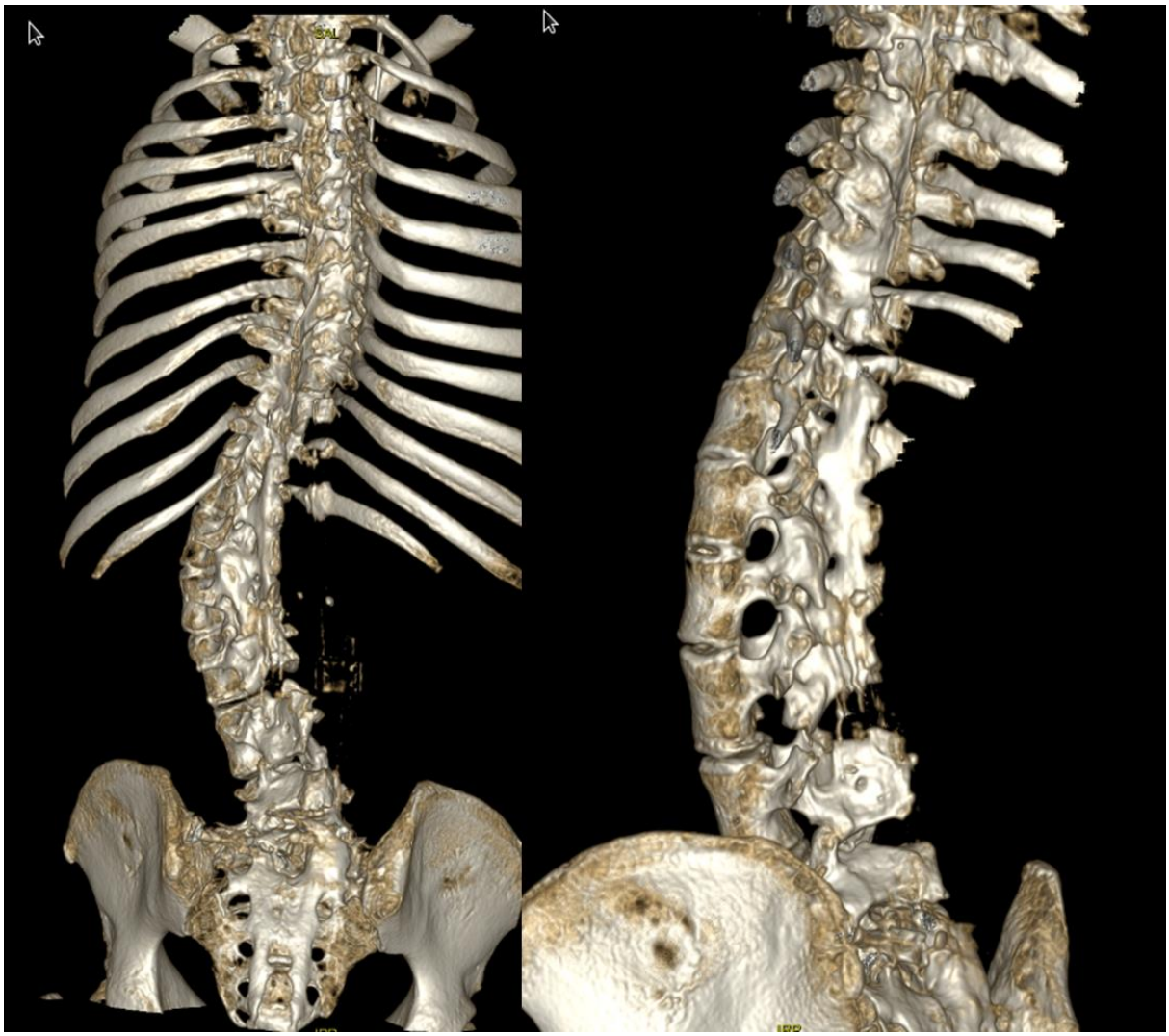

Figure 3 :3D reconstruction. The rods have been removed to allow a better view of the ossifications.

Blind operators agreed on 98% cases of facet joints fusion and 96% of ossifications around the rods.

Mean preoperative and last follow-up thoracic vertebrae height were respectively 13.46mm (SD = 1.75) and 17.4mm (SD = 1.7) with a mean difference of -4.0 (SD = 1.8; CI95% = -5.2; -2.7; $p<0.001$. Mean preoperative and last follow-up thoracic intervertebral disc height were respectively 3.6mm (SD = 0.29) and 2.05mm (SD = 0.5) with a mean difference of -1.58 (SD = 0.6; CI95% = 1.1; 2; $p<0.001$. Mean last follow-up of lumbar vertebrae height was 26±2mm. (Figure 4).

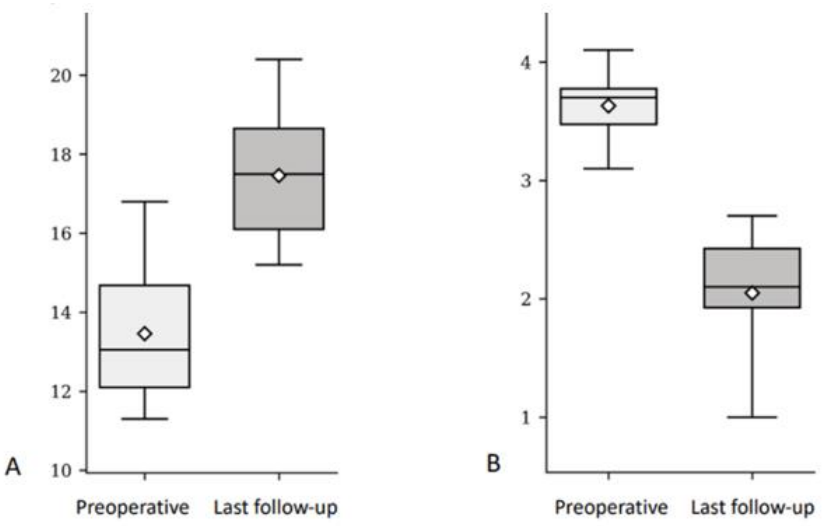

Figure 4: (A) Thoracic vertebral height, (B) Thoracic intervertebral disc height.

Vertebral growth can be evaluated when comparing the postoperative and preoperative CT scans. Preoperatively, only thoracic CT scan were available apart from one patient to whom a TAP CT-scan was performed. Thus, postoperative lumbar height was not available (Figure 5).

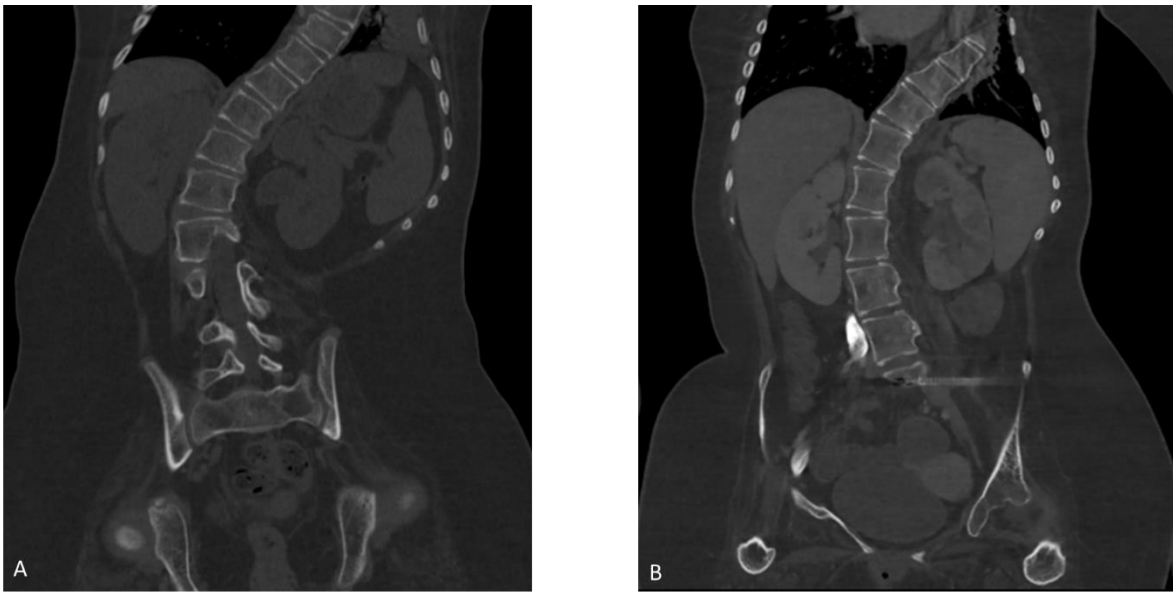

Figure 5 : A/Preoperative and B/ after 8 years follow-up CT scan of an SMA type 2 patient showing an increase of the thoracic and lumbar vertebral height, and a decrease of discal height.

DISCUSSION

The present computed tomography study is the first to describe and quantify the rate of fusion at the end of MIFBF surgery program. The fusion of facet joints and the induced bone around the rod suggest that the level of autofusion which was spontaneously obtained could justify the avoidance of PSF. The analysis of complication rates at medium-long term (6.5 years [18]) corroborates this interpretation. These results may radically change our surgical guidelines for patients with neuromuscular scoliosis.

MIFBF was initially used as an alternative to traditional growing rods to reduce mechanical and infectious complications while waiting for PSF, which was initially planned at skeletal maturity for all patients. However, stable clinical and radiological correction was observed with a reduced rate of mechanical complications [18, 20]. Fibrosis and spontaneous fusion around the implants and along the rods were observed during late surgeries performed in some patients and had confirmed the absence of the need to perform PSF as initially planned (fig 1).

The literature on the physiopathology of such progressive autofusion of the spine is poor. Autofusion was first described in 1984, by Moe et al. [28] who reported spontaneous fusion at the curve extremities and/or at the curve apex in young children treated for severe deformities with subcutaneous Harrington rod without fusion. Mardjetko et al [29] observed extensive fibrosis and laminar spontaneous fusions under the instrumented regions with Luqué rods, frequently involving the entire thoracic spine but also extending across the thoracolumbar spine. Cahill et al. [30] reported that early autofusion of the spine, can be caused by a combination of repeated surgical procedures and spontaneous bone bridging in the immobilized spinal span, and occurs in as many as 89% of children treated with growing rods. Early autofusion may also result in diminishing amounts of lengthening, also called 'law of diminishing returns' achieved

after rod lengthening as well as the reduced ability to gain deformity correction at the final definitive fusion surgery [31,32].

In the present study, the autofusion after MIFBF could be related to the increased rigidity of the spine provided by the strong and stable bipolar construct, anchored proximally by a double hooks' claws and distally by ilio-sacral screws. However, thanks to the minimally invasive approach, the preservation of the intermediate area between the proximal and distal fixation avoided too early autofusion that could have led to cessation of growth like in traditional growing rods techniques. Thus, thoracic and lumbar vertebral height at skeletal maturity achieved the corridor of normal height (Fig 2) [26].

In opposition with traditional growing rod techniques, where the lengthening connector is generally inserted in the intermediate area leading to early fibrosis and autofusion due to repeated surgical lengthening procedures at this area, the bipolar fixation provides delayed autofusion that happens progressively many years later. This delayed autofusion in the bipolar technique allows to improve the correction of the residual deformity thanks to repetitive rod lengthening or to the use of expandable devices in the first years after index surgery.

That may explain why in most of cases the amount of spinal and pelvic deformity correction obtained by the bipolar technique was higher than with other growing rod techniques and even with arthrodesis in this population of patients [33,34].

In the bipolar technique, the early autofusion may exists proximally in the instrumented thoracic vertebrae and distally in the lumbosacral junction secondary to the surgical approach and the insertion of the implants. The presence of metal close to the bone at the lumbosacral junction induces a progressive autofusion that cover the implants and stabilize them.

However, the amount of autofusion and its time to onset may vary from patient to patient. This study is limited by the small number of patients who underwent a full spine CT scan. The

answer how to act after the end of growth in this patient group is far from solved: to remove or to leave the implants in situ [35], to make a rod lengthening or to make a reoperation with supplementary correction and instrumented fusion [36,37]. However, in view of the results of this study, and the low frequency of neuromuscular scoliosis, we may recommend performing a low dose CT scan before any clinical decision at final lengthening program to confirm the no need of final PSF [38]. Additional studies will be necessary to better understand the progressive stiffening of the spine on surrounding tissues.

CONCLUSION

The present study suggests that final fusion after minimally invasive fusionless bipolar fixation might be unnecessary. This hypothesis is supported by the evidence of a high rate of spontaneous fusion on CT-scan at the end of the lengthening program in this series of neuromuscular patients. Further investigation on a larger cohort should confirm these findings. Furthermore, the impact of this less invasive surgical technique and of the stability obtained should be analyzed in view of long-term complications and quality of life of the patients.

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Cette première étude quantitative a pu montrer que les articulaires postérieures étaient fusionnées à 93% en moyenne, et que la croissance vertébrale avait été préservée chez l'ensemble des patients. Cependant, le faible nombre de patients inclus n'a pas permis d'évaluer des facteurs favorisants la fusion spontanée.

Dans le chapitre suivant, nous avons voulu évaluer si d'autres facteurs anatomiques contribuent à l'enraidissement rachidien, en particulier les disques intervertébraux, grâce à la réalisation d'élastographies discales.

7. Analyse discale par élastographie de l'annulus fibrosus lombaire

7.1. Article 4 : Evidence of Spinal Stiffening following Fusionless Bipolar Fixation for Neuromuscular Scoliosis. A Shear Wave Elastography Assessment of Lumbar Annulus Fibrosus.

M Gaume, K Loiselet, H Chekir, T Langlais, N Boddaert, S Pannier, W Skalli, L Miladi, C Vergari

Résumé

Objectif

Il n'existe pas de critères établis pour quantifier l'enraidissement rachidien après instrumentation sans greffe bipolaire chez les patients atteints de scoliose neuromusculaire (SNM). Il n'y a pas non plus de consensus sur la stratégie chirurgicale à proposer à long terme. Cette étude rapporte la première utilisation de l'élastographie par ondes de cisaillement pour évaluer la réponse mécanique des disques intervertébraux lombaires (DIV) après instrumentation sans greffe, et comparé à des sujets témoins sains. L'objectif était d'obtenir des preuves de la rigidité de la colonne vertébrale après instrumentation sans greffe.

Méthodes

Dix-neuf SNMS opérés par instrumentation sans greffe bipolaire (18 SD2 ans au dernier recul, 6,2 SD1an après la chirurgie initiale) ont été inclus prospectivement. L'angle de Cobb préopératoire était de 89SD20° et de 35SD1° au dernier recul. Tous les patients avaient atteint la maturité squelettique. Dix-huit patients sains (20±4 ans) ont également été inclus. La vitesse de l'onde de cisaillement (SWS) a été mesurée dans l'anneau fibreux des DIV L3L4, L4L5 et L5S1 et a été comparée entre les deux groupes. La fiabilité des mesures a été vérifiée.

Résultats

Chez les sujets sains, le SWS moyen (tous niveaux discaux confondus) était de 7,5SD2,6m/s. Chez les patients atteints de SMN, le SWS était significativement plus élevé, à 9,9 SD 1,4m/s ($p < 0,05$). Les différences étaient significatives entre L3L4 (9,3SD1,8m/s vs 7,0SD2,5m/s, $p=0,004$) et L4L5 (10,3SD2,3m/s vs 7,1SD1,1m/s, $p=0,0006$). Aucune différence n'a été observée pour L5S1 ($p=0,2$). Aucune corrélation n'a été trouvée avec l'âge au moment de la chirurgie, ni avec la correction de l'angle de Cobb et l'âge au moment de la mesure.

Conclusions

Cette étude montre une augmentation significative de la rigidité discale en fin de croissance chez les patients atteints de NMS traités par FBF. Ces résultats pourraient être un complément intéressant à la tomodensitométrie pour évaluer la rigidité de la colonne vertébrale, permettant d'éviter une arthrodèse vertébrale postérieure à la maturité du squelette.

Objective

There are no established criteria for stiffness after fusionless surgery for neuromuscular scoliosis (NMS). As a result, there are no consensus regarding the surgical strategy to propose

at long-term follow-up. This study reports the first use of shear wave elastography for assessing the mechanical response of lumbar intervertebral discs (IVDs) after fusionless bipolar fixation (FBF) for NMS and compare them with healthy controls. The aim was to acquire evidence from the stiffness of the spine following FBF.

Methods

Nineteen NMS operated on with FBF (18SD2y at last follow-up, 6.2SD1y after surgery) were included prospectively. Preoperative Cobb was 89SD20° and 35SD1° at latest follow-up. All patients had reached skeletal maturity. Eighteen healthy patients (20±4y) were also included. Shear wave speed (SWS) was measured in the annulus fibrosus of L3L4, L4L5 and L5S1 IVDs and was compared between the two groups. A measurement reliability was performed.

Results

In healthy subjects, average SWS (all disc levels pooled) was 7.5SD2.6m/s. In NMS patients, SWS was significantly higher at 9.9 SD 1.4m/s ($p < 0.05$). Differences were significant between L3L4 (9.3SD1.8m/s vs 7.0SD2.5m/s, $p=0.004$) and L4L5 (10.3SD2.3m/s vs 7.1SD1.1m/s, $p=0.0006$). No difference was observed for L5S1 ($p=0.2$). No correlation was found with age at surgery, nor with Cobb angle correction and age at the SWE measurement.

Conclusions

This study shows a significant increase of disc stiffness at the end of growth for NMS patients treated by FBF. These findings are a useful adjunct to CT scan in assessing stiffness of the spine allowing the avoidance of surgical final fusion at skeletal maturity.

INTRODUCTION

Neuromuscular scoliosis (NMS) is due to a lack of muscular control caused by a neurological or degenerative muscular condition. When conservative treatment by bracing fails to control curve progression, patients may require surgery at an early age. In such situations, different growth-friendly techniques have been described [1]. These fusionless techniques allow deformity correction while preserving spinal and thoracic growth. At skeletal maturity, which corresponds to the endpoint of the lengthening protocol, or even frequently before, a definitive posterior spinal fusion (PSF) is usually performed. PSF consists in fusing all vertebral levels with a posterior metallic instrumentation that stiffens the spine and stops almost any intervertebral movement. However, PSF stops also spinal and thoracic growth, and it is at high risk of complications, including hemorrhage [2], infections and mechanical complications [3]. Our team used a bilateral fusionless bipolar fixation (FBF), which became an alternative

technique for PSF in NMS scoliosis in our department. This technique is based on a telescopic construct including two 5.5 mm diameter rods on each side bridging the curve and linking two proximal-distal anchors. It is a modular construct which follows spinal growth and allows further gradual correction of residual deformities. The minimally invasive approach minimizes the risk of early fibrosis and auto-fusion, allowing the telescopic construct lengthening for longtime to preserve growth until skeletal maturity. Over the long term with a follow-up of more than 10 years for the first cases, a progressive stiffening was supposed by the continued stability of the spinal correction at skeletal maturity, suggesting that PSF was unnecessary[4,5]. In other words, clinical observations suggested that a natural stiffening of the spine was occurring, which could make spinal fusion by instrumentation superfluous.

However, evidence of spinal stiffening following FBF for NMS has never been analyzed. It was hypothesized that the origin of this stiffening could be i) a peri-implant bone growth, ii) bone growth within the disc, or iii) a general stiffening of the disc, possibly due to partial mineralization. The present study aims to elucidate this last aspect.

Bony structures of the spine are typically evaluated using computed tomography (CT), while magnetic resonance imaging (MRI) is used to assess soft tissue. MRI has been used to assess disc stiffness, its access is difficult in clinical routine, and it requires a long period in an uncomfortable position, especially in neuromuscular patients.

Ultrasound imaging modalities are non-invasive, known for their rapidity, safety and portability. Particularly, shear wave elastography (SWE) is an imaging technique based on the propagation velocity of the ultrasonic induced shear wave to quantify the soft tissue elasticity or stiffness [6,7]. SWS reflects the intrinsic mechanical properties of the tissue, and a higher SWS is associated to a stiffer tissue [8]. It has been previously applied to evaluate IVD in adolescent idiopathic scoliosis, and it showed that IVDS in patients had higher shear wave

speed (SWS, i.e., stiffer tissue) than asymptomatic controls [9,10]. Furthermore, it was shown that this increased SWE can detect changes in IVD, and in particular disc SWS in idiopathic scoliosis tended to normalize one year after fusion surgery[11].

This study reports the first use of SWE for assessing the mechanical properties of IVD in NMS after FBF and compare them with asymptomatic controls. The aim was to acquire evidence from the stiffness of the spine following FBF at long term follow-up.

MATERIAL AND METHODS

Subjects

Data were collected prospectively from our department within follow-up clinical routine, between January 2021 and June 2022. Parents and children were informed about the measurement protocol and consented to participating before inclusion. Parents signed informed consent, which was approved by the ethics committee (IDRCB 2020-A03598-31, ClinicalTrials.gov Identifier: NCT04969770).

Nineteen NMS patients who underwent FBF from T1 to the pelvis were included prospectively in this study (7 girls and 12 boys) . Preoperative Cobb angle was 89°±20,and 35°±1 at last follow-up. They were 11±2 years old at initial surgery and 18±2 years old when SWE was performed. Twelve had cerebral palsy, six were syndromic, and one suffered from spinal muscular atrophy type 2. All were non ambulatory and had reached skeletal maturity at last follow-up. Patients with less than 5 years postoperative follow-up were excluded.

Eighteen asymptomatic subjects were also included as control group (9 girls and 9 boys, mean age 20±3y). Healthy children were included if they had no history of spinal disease, and a systematic clinical examination was performed by a spine surgeon to rule out any diagnosis of scoliosis.

Protocol

Shear-wave speed (SWS) was measured in all included subjects following a standardized protocol determined for lumbar IVD in children and adolescents' idiopathic scoliosis [12].

Patients were in supine position and the probe was placed on the abdomen to image the spine with an anterior approach. The thinness of the abdominal wall in children allows a good quality ultrasound exploration, which is much easier than in adults. It should also be noted that in the operated group, the scoliosis brings the lumbar spine forward, and therefore even closer to the anterior abdominal wall.

Measurements were obtained using a Canon Aplio i800 system[13]. The probe we used was the i8CX1 (or PVI 475BX) which is a convex probe with a frequency range between 1 and 8 MHz (Mode Multi, Range 0-75KPa). The probe was pushed to 2–3 cm from the disc. The aortic bifurcation was localized to determine the L4 vertebral level.

Ultrasound examination started with a sagittal section under the umbilicus, which allowed to locate the vertebral bodies and the intervertebral discs. The vertebrae are identified by a very hyperechoic linear interface with a posterior shadow cone, in connection with the cortical anterior wall (Figure 1). The discs are hypoechoic and allow ultrasound to pass posteriorly.

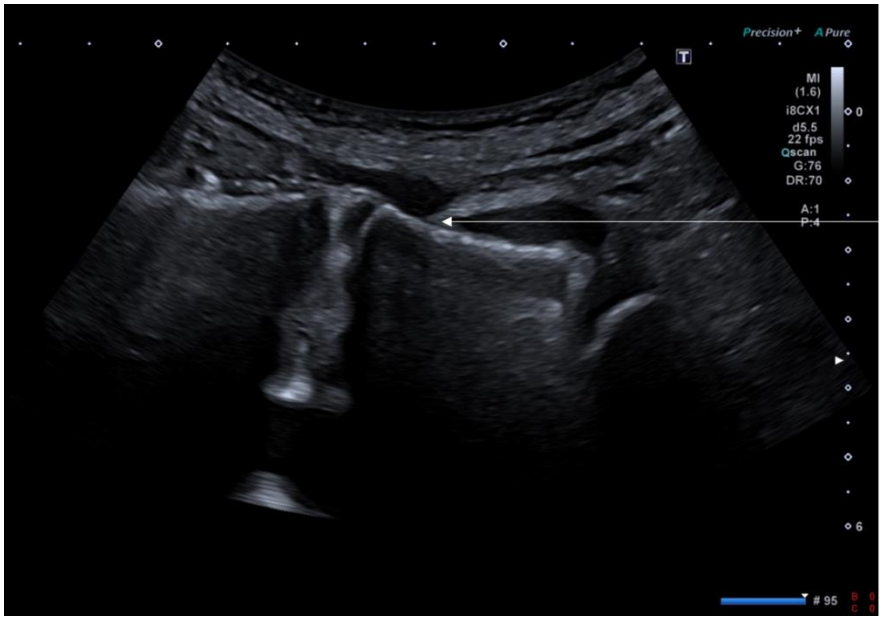

Figure 1 : Sagittal ultrasound section. V = vertebrae (V); IVD = intervertebral discs

After a 90° rotation of the probe, the operator switched in axial section in the plane of an intervertebral disc. The medullary canal with the roots can be seen behind the disc. The posterior joints are also visible laterally (Figure 2). The probe was subsequently moved up or down to measure SWS in the annulus fibrosus of L3-L4, L4-L5 and L5-S1 IVDs [14]. For L5-S1 disc, a greater inclination of the probe was necessary to be in the plane of the disc. Children with a transitional anomaly were excluded from both groups.

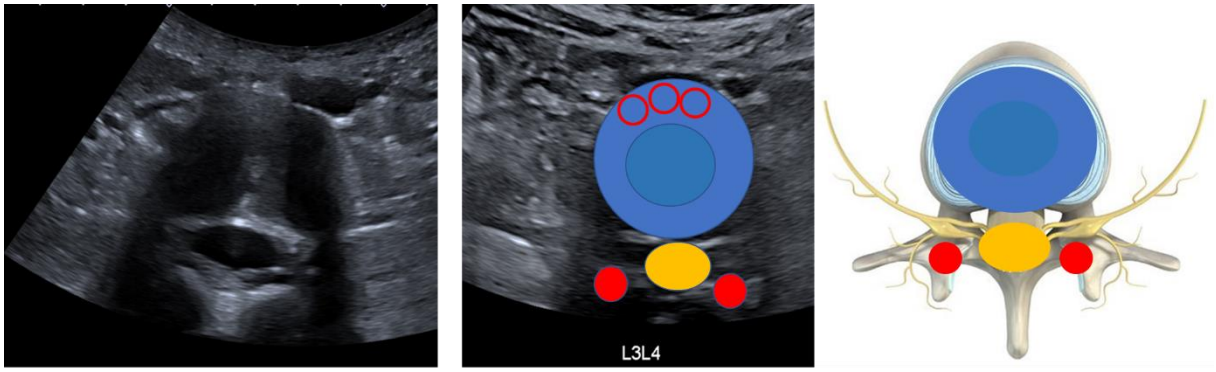

Figure 2 :Axial ultrasound section in the plane of an intervertebral disc.

The medullary canal with the roots can be seen behind the disc. The posterior joints are also visible laterally.

Disc in blue, spinal canal in yellow, post articular in red. Red circles: Area of interest in the annulus fibrosus.

For each IVDs (L3-L4, L4-L5 and L5-S1), 3 circular regions of interest (ROIs) of 8mm diameter were defined in the periphery of the intervertebral disc at the annulus (Figure 3) .This procedure was repeated three times, and the average was calculated to obtain a single SWS value.

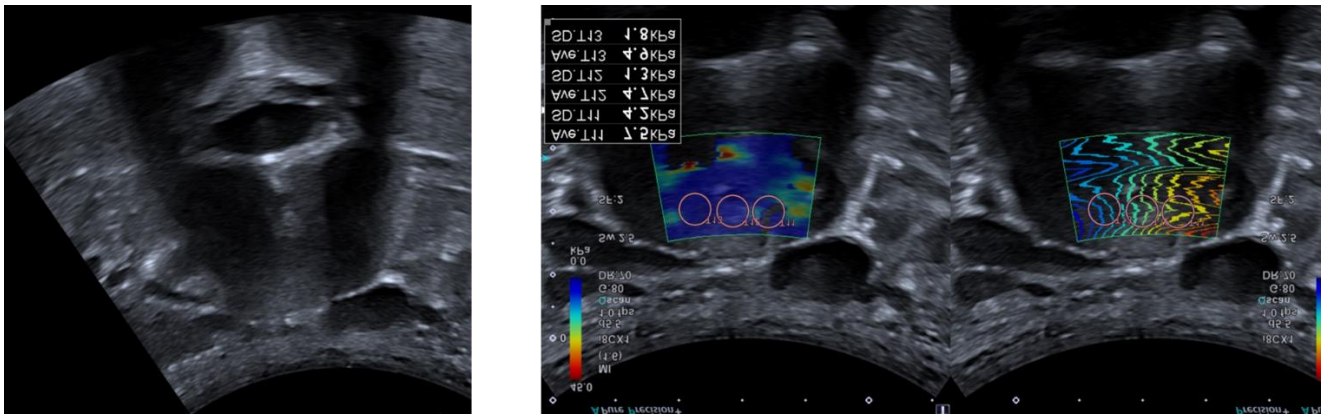

Figure 3 : Ultrasound and corresponding elastogram with3 circular regions of interest defined in the periphery of the intervertebral disc at the annulus.

Measurement reliability was assessed on a subset of the first five NMS and healthy patients by two confirmed radiologists. Then, only one of them according to his availability performed the subsequent measures.

Statistics

Intra-operator repeatability and inter-operator reproducibility were determined according to ISO 5725 standard[15]. Five sets of three measurements were repeated by two trained radiologists. The probe was repositioned after each set of measurements. Repeatability and reproducibility were calculated in terms of standard deviation of uncertainty. Intraclass correlation coefficient (ICC) was determined both intra- and inter-operator; an ICC greater than 0.75 was considered to signify good agreement.

Differences were analyzed with Wilcoxon rank sum tests and correlations were quantified with Spearman's rank correlation coefficient, since variable did not follow normal distributions (Shapiro Wilk test).

Differences between NMS and healthy groups were analyzed with Mann-Whitney tests and differences between vertebral levels with Kruskal-Wallis tests. Correlations were quantified with Spearman's rank correlation coefficient.

Significance was $p<0.05$. Analyses were performed using Microsoft Excel 2016.

RESULTS

Intra-operator repeatability was 4.3% and inter-operator reproducibility was 5.2%. Intraclass correlation coefficient was higher than 0.8 for each operator, showing "good agreement" for NMS patients and healthy patients (mean ICC=0.89 and 0.91, respectively).

On average, the investigation lasted less than 30 minutes per patient, including the installation from the wheelchair to the examination table.

Figure 4 reports SWS in annulus fibrosus at the three-disc levels for the two groups (asymptomatic and NMS).

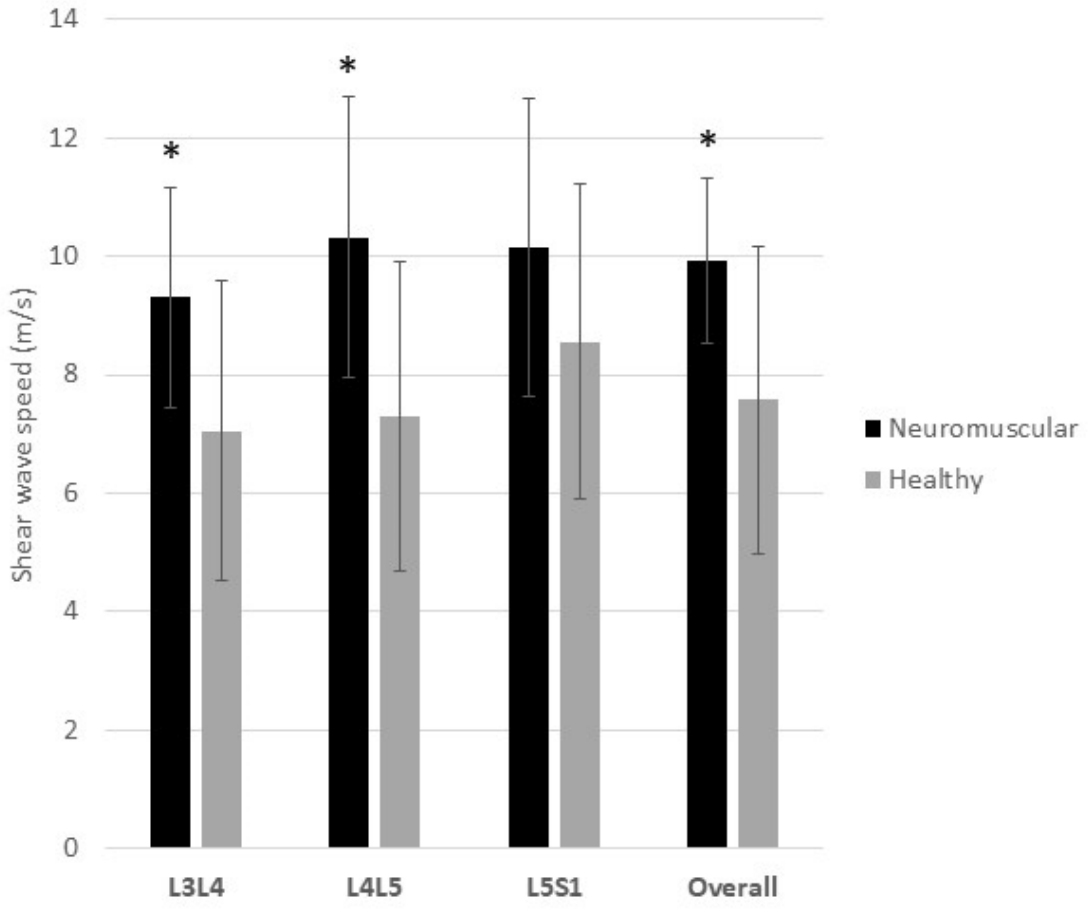

Figure 4 :Shear wave speed comparison in annulus fibrosus at the three-disc levels for the two groups.

Healthy group SWS

There was no significant difference between levels L3-L4 (7.05 ± 1.3m/s), L4-L5 (7.3 ± 1.1m/s) and L5-S1 (8.5±1.6m/s, $p > 0.05$), Figure 4), so values were pooled to obtain an overall average of 7.5 ± 2.6m/s . The difference in average SWS between girls and boys was not significant (p= 0.2). No correlation of SWS was found with age.

NMS group SWS

There was no significant difference between levels L3-L4 (9.3±1.8 m/s), L4-L5 (10.3±2.3m/s) and L5-S1 (10.15±2.5m/s, $p > 0.05$, Figure 4), and overall average SWS was 9.9±1.4m/s. No correlation was observed with age at surgery, gender, age at the SWE measurement, time between surgery and SWE measurement, and Cobb angle correction.

Comparison between groups

Differences between NMS and healthy subjects was significant at L3L4 ($p=0.004$) and L4L5 disc levels ($p= 0.0006$), but not at L5S1 ($p=0.3$). Difference between overall averages was significant ($p=5.627E-6$)

DISCUSSION

The avoidance of PSF at the end of lengthening program of growth guidance techniques in NMS has been recently an active area of research. PSF is indeed often regarded by patients as the conclusion of a long burden. Poe-Kochert et al. [16] reported 20% of unplanned reoperation after PSF following traditional growing rods program.

In a series of 100 NMS treated by FBF [17], PSF initially planned at skeletal maturity was not performed for any patients. PSF was avoided according to the stable correction of spinal and pelvic deformities observed during follow-up, and by the strength of this type of fixation [19,18]. This stability over time suggested a progressive spinal stiffening following FBF. Thus, the spinal growth was possible in the first years of surgery, and the gradual increase of spinal stiffening made it possible to maintain the correction over time.

However, evidence of spinal stiffening following FBF for NMS has never been analyzed. In particular, the role of the intervertebral disc (IVD) in this stiffening process is not yet clear.

In this paper, SWE was used to obtain information on lumbar IVD mechanical properties between 5 and 8.1 years after surgery, to shed some light on spinal stiffening after FBF in NMS.

This recent imaging procedure could be particularly attractive for NMS patients, who have difficulties to lie down for a long time in an CT scan or magnetic resonance imaging, because of the spinal deformity or abnormal movements related to their pathology . Overall, measurement time was much shorter than a typical CT scan, and more comfortable for the patient. The measurement reliability indicated that it is feasible in this population with intra-operator repeatability of 4.3 % and inter-operator reproducibility of 5.2 %.

Until now, SWE has been used in NMS patients for the study of paravertebral muscles [20,21] but has never been applied to lumbar IVD, which is already well studied in patients with adolescent idiopathic scoliosis (AIS). Langlais et al. [9] showed that AIS presented a stiffer annulus fibrosus than asymptomatic adolescents, with SWSs of 3.5±0.3 m/s vs 3.0±0.3 m/s, respectively. Values obtained in this study were much higher, which is probably due to the different technologies employed in the two studies (Canon Aplio i800 system in the present series versus Aixplorer Supersonic Imagine).

Therefore, it is important to keep in mind that SWS disc measurement can be very sensitive to the device that is used, and comparison to values obtained with different machines is not relevant, even if the measurement with each machine appears reliable, the technical differences induce a bias of comparison.

However, it can be noted that idiopathic scoliotic patients with no surgery showed a SWS 1.2 times higher than controls, while SWS in this study was 1.3 times higher than controls. This difference is indicative of the fact that idiopathic scoliosis patients present stiff but mobile spine, while NMS patients show stiffer spine, which raises the question of some degree of autofusion. Besides, a complete ossification of the disc would have made the measurement impossible, since the elastographic signal would not be obtained in bone (Figure 5).

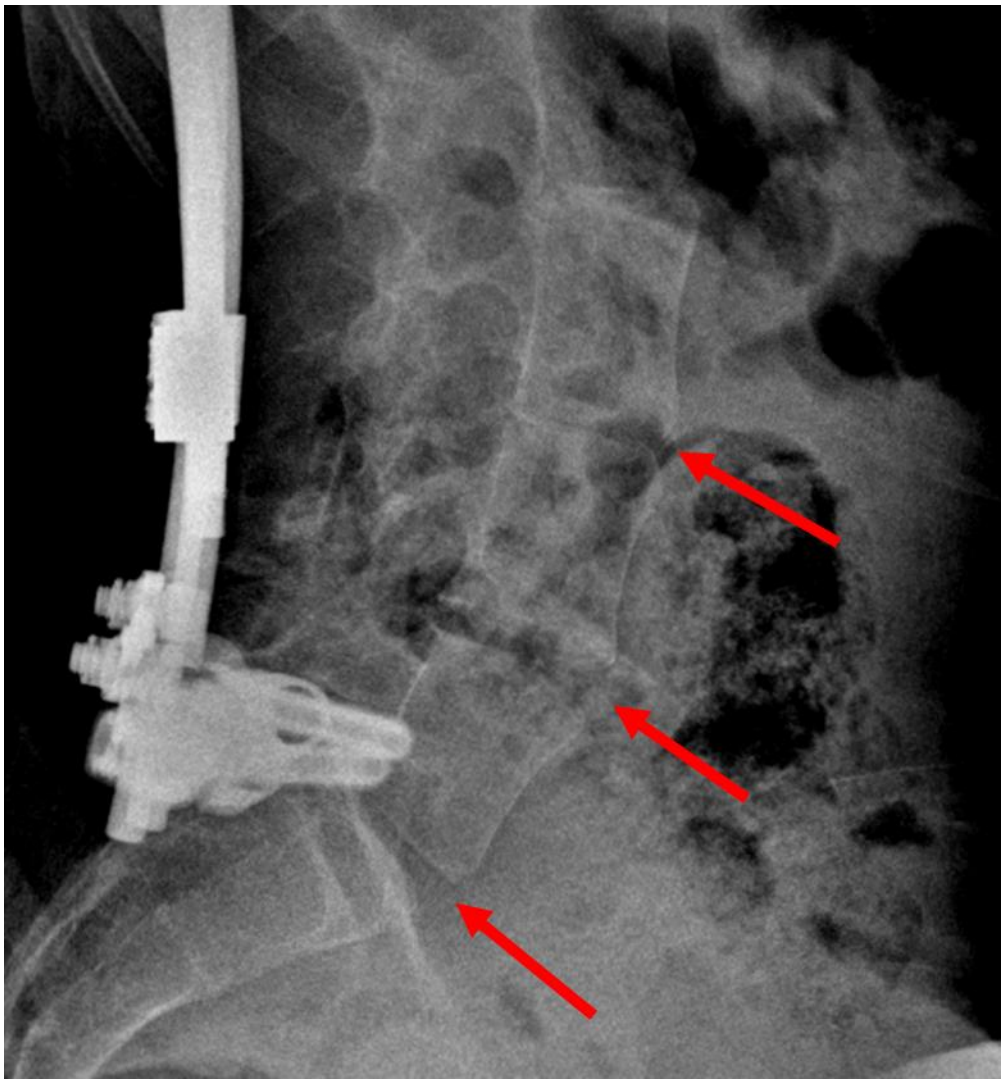

Figure 5 : The absence of discal ossification permitted to obtain an elastographic signal through the anterior approach.

Unlike previous work on IVD ultrasonography and elastography, microstructural features of the disc were not visible in the present work. This is probably due to the use of a low-frequency probe, which resulted in a spatial resolution too low to distinguish the lamellar structure of the annulus. Assuming an average frequency of 3 MHz and a speed of sound in the annulus fibrosus of 1600 m/s, the spatial resolution should be about 0.5mm, whereas lamellae in adolescents have a thickness in the order of 0.2 mm.[22]

Contrary to Deviren et al. [23], no significant positive correlation was found between the spinal curvature severity and the stiffening of the spine. Vergari et al.[14] compared SWE in AIS before and after surgical intervention, showing that discs outside the fusion tended to normalize one year post fusion surgery. This is not in contradiction with the present results, since the discs analyzed in this study were within the instrumentation, and therefore they could not recover their flexibility.

One of the limitations of this study was that measurements were performed with an anterior approach, which does not allow accessing the thoracic discs. However, posterior approach is impossible due to the presence of the bony neural arch, and mediolateral approach is difficult because of the larger depth of the disc in this approach. The other limitation is the possible bias in the measurement caused by the modification of the stiffness according to the posture [24,25]. The stiffness of lumbar IVDs in lying position during the imaging may be underestimated compared to sitting posture on wheelchair. However, this bias was reduced because asymptomatic controls were assessed in the same posture. Finally, machine difference can be an obstacle for this application to be standardized. However, it could be possible to normalize standardizing the measurement, for example, by doing a measuring soft-tissue SWE at a specific location as the baseline and use this to normalize the SWE value.

CONCLUSION

In this study we present the first attempt to quantify the stiffness of lumbar discs in NMS, compared to healthy patients, using SWE. This technique proved to be a feasible, fast, and reliable means of quantifying disc mechanical properties and stiffening of the spine after FBF. This could be is an additional argument when questionning the real need for PSF at skeletal maturity.

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Cette deuxième étude quantitative a pu montrer que l'enraidissement rachidien au cours de l'instrumentation sans greffe avait également une origine discale. Cette technique de quantification s'est avérée fiable, rapide et non irradiante. Il pourrait donc d'agir d'un examen utile d'évaluation de l'enraidissement au cours du suivi, avant la réalisation d'un scanner low-dose, qui reste irradiant.

Dans le chapitre suivant, nous avons voulu évaluer, après les propriétés osseuses et discales, les qualités musculaires du rachis instrumenté à long terme, grâce à la réalisation d'IRM musculaires.

8. Analyse musculaire par résonnance magnétique.

8.1. Article 5 : Thigh and paraspinal muscles changes after fusionless bipolar fixation for early onset scoliosis in type 2 spinal muscular atrophy

M. Gaume, C Vergari, M Creze, A Bonnet-Lebrun, C Muth-Seng, E Ferrero, S Pannier, S Quijano-Roy, L Miladi, W Skalli, R Carlier.

Résumé

Introduction. L'objectif de cette étude prospective était de mieux comprendre les modifications des muscles de la colonne vertébrale et de la cuisse chez des sujets atteints d'amyotrophie spinale de type 2 (ASI2), avant et après une chirurgie mini-invasive par instrumentation sans greffe.

Méthodes et patients. 10 patients avec confirmation génétique de SMA2 et présentant une scoliose neuromusculaire sur les radiographies bi-planaires à faible dose ont été inclus : 5 patients préopératoire, et 5 patients postopératoire qui avaient un recul minimal de 5 ans après la chirurgie. La chirurgie a consisté en une instrumentation sans greffe bipolaire. Il a été réalisé chez tous les sujets une imagerie par résonance magnétique musculaire avec séparation graisse/eau du rachis thoracique à la cuisse. Le pourcentage de dégénérescence graisseuse a été comparé avant et après chirurgie. Une enquête de qualité de vie a été réalisée.

Résultats. L'infiltration graisseuse était diffuse et symétrique dans les deux groupes de patients, et en moyenne six fois plus importante par rapport aux sujets témoins précédemment publiés au niveau de la cuisse. Les adducteurs, le sartorius et le gracilis étaient moins touchés avec respectivement 51%, 56% et 57% de fraction graisseuse avant la chirurgie. En comparant les groupes préopératoires et postopératoires, l'infiltration graisseuse était plus importante dans le sartorius et le multifidus après chirurgie ($p<0,05$). Aucune différence significative n'a été retrouvée pour les autres muscles étudiés. Ces résultats n'ont pas affecté la qualité de vie, qui est apparue globalement améliorée.

Conclusion. Il s'agit de la première étude comparant l'infiltration graisseuse des muscles para spinaux et des muscles de la cuisse chez les patients atteints d'ASI2 avant et après une chirurgie de type instrumentation sans greffe bipolaire. Nos résultats montrent que les muscles sont globalement préservés après la chirurgie en dehors du multifidus et du sartorius.

Abstract

Introduction. The aim of this prospective study was to better understand modifications in spinal and thigh muscles of subjects with type 2 spinal muscular atrophy (SMA2) and early onset scoliosis, before and after minimally invasive fusionless surgery.

Methods and Patients. 10 genetically confirmed SMA2 patients with confirmed scoliosis on bi-planar low-dose X-ray were included : 5 preoperative and 5 postoperative patients with a minimal follow-up of 5 years after surgery. The surgery consisted in a bilateral sliding rods construct extended from T1 to the sacrum, anchored proximally by double-hook claws and

distally by ilio-sacral screws through a minimally invasive approach. All subjects had a fat/water separation muscle magnetic resonance imaging from spine to the thigh. The percentage of fat degeneration was compared before and after surgery. A quality-of-life survey was performed.

Results. Fat infiltration was diffuse and symmetric in both groups of patients, and on average six time more compared to control subjects previously published at thigh level. Adductors, sartorius and gracilis were less affected with respectively, 51%, 56% and 57% of fat fraction before surgery. Comparing the preoperative and postoperative groups, fat infiltration was higher in sartorius and multifidus after surgery ($p<0.05$). No significant difference was found for the other muscles studied. These results did not affect quality of life which appeared globally improved.

Conclusion. This is the first study to compare fat infiltration of spinal and thigh muscles of SMA2 patients before and after minimally invasive surgery. Our results demonstrate that muscles were globally preserved after surgery apart from multifidus and sartorius that were more affected.

INTRODUCTION

Spinal muscular atrophy (SMA) is a severe, progressive autosomal recessive neuromuscular disorder in which there is a degeneration of anterior horn cells associated with progressive loss of motor function [1-5]. Patients with type 2 SMA develop their symptoms between the ages of 6 and 18 months, with progressive proximal muscle weakness and mild to moderate hypotonia. Patients acquire sitting but they are unable to walk. Neuromuscular scoliosis prevalence reaches 67% in this population and starts from very early ages [6]. When bracing is not sufficient to control the deformity, early spinal surgery is required [7]. Fusionless surgical techniques allow correction of the deformity, prevent its progression and preserve spinal and thoracic growth [8]. Magnetic resonance imaging (MRI) provides a high soft tissue contrast allowing assessment of healthy striated muscles concerning shape, volume, and tissue architecture. It also allows detection of increase of fat infiltration into muscles. As a radiation-free modality, it is a valuable noninvasive imaging method in children. Its use has been increasing to diagnose, to characterize the severity and to monitor progression of muscular disorders [9,10]. Few studies have reported muscle involvement using MRI in SMA2 patients. At thigh, pattern of selective muscle involvement has been reported, such as the adductor longus and gracilis that are always

selectively spared [11]. However, to the best of our knowledge, no work in the literature have assessed modifications in spinal and thigh muscles of subjects with SMA2 following spinal surgery. The aim of the present study was to better understand modifications in fat infiltration following such surgery.

METHODS

Patient population

This was a single center study, conducted prospectively between 2021 and 2023. We included 10 genetically confirmed SMA2 patient, 5 preoperative (4 female, 1 male , aged 11.3y , range 10.0 -12.2y), and 5 postoperative patients with a minimum of 5 years follow-up (5 female, aged 11.6y, range 9.8-13.1y at initial surgery, and aged 18.1, range 16.3-20y at the day the MRI was performed). All were sitters but non-ambulatory. Exclusion criteria included patients with tracheostomy or severe respiratory problems, patients with abnormal movements, major contracture, presence of pacemakers, need for sedation and claustrophobia. Clinically relevant data were obtained from available medical records. Patients were assessed using a quality-of-life survey, including back pain (yes/no. If yes, measurement on a scale from 0 to 10) , ability for transfers (better/worse/the same) and satisfaction after surgery (yes/no). Parental/legal guardian written informed consent was obtained prior to inclusion**.** The study was approved by the appropriate ethics committee (CPP, RIPH-3, RCB 2020-A02985-34).

Surgical technique

The minimally invasive fusionless bipolar fixation technique was previously described [12]. Two small midline skin incisions are made: one proximal at the upper thoracic spine and one distal at the lumbosacral junction. The fixation extends from T1 to the sacrum proximally anchored with hook claws [13] and distally by iliosacral screws [14,15]. Rod lengthening

procedures thought the distal incision are proposed to achieve further correction in case of residual spinal deformity or in case of curve worsening because of growth spurt.

Protocol and measure

All patients underwent head to thigh muscle MRI on a 3-T General Electric MRI system (Discovery™ MR750w, General Electric Medical Systems, Milwaukee, United States of America), with a network of phased array coils . The technique required precise patient positioning in supine position with upper limbs lying close alongside the body. Braces fitted to the table ensured patient safety. No contrast material was administered. The axial slices were consecutive, parallel, and contiguous with a constant slice thickness of 5 mm. The imaging protocol was the Dixon 2-points protocol [20], first set of images where the intensity of each voxel was correlated with the amount of water (Water image) and a second set of images where the intensity of each voxel was correlated with the amount of fat (Fat image) were automatically generate. These two sequences had the same positions and orientations of the slices.

All MRI data were processed using custom software [19-20] in a total of 15 muscles groups (latissimus dorsi, erectus spinae, psoas, iliacus, gluteus maximus, gluteus minimus, medius and tensor fasciae latae, sartorius, rectus femoris, musculus vastus, biceps femoris, semitendinosus, semimembranosus, gracilis, adductor and pelvi-trochanteric muscles). The fatty infiltration (FI) was evaluated by a calibration between a purely fatty subcutaneous area (100% FI) and a purely muscular area in the psoas (0% FI).

Measurements were assessed by one experienced musculoskeletal radiologist and one orthopedic surgeon who had experience of examining muscle images of neuromuscular disorders. In case of discrepancy between examiners, a consensus was reached.

Statistical analysis

Statistical analyses were performed using SPSS Statistics software (version 23.0; IBM). A descriptive analysis of demographic data was performed (N, mean, SD). Fat infiltration was compared between groups with a Mann-Whitney test. The level of significance was set at $p<0.05$.

RESULTS

Patient population

Of the 10 preoperative patients included, 5 did not complete the MRI acquisitions because of pain in lying position, panic, or stress despite psychological preparation. Of the 10 postoperative patients included, 5 patients had initially accepted but finally refused because of the additional burden of the research protocol. There were no significant differences in terms of age at initial surgery, preoperative Cobb angle and pelvic obliquity between the two groups. Clinical and radiological characteristics of included post perative patients are shown in Table 1.

Fat infiltration pattern

Fat infiltration comparison between muscles is summarized in Figure 1 and Figure 2.

Fat infiltration was diffuse and symmetric in all patients pre and postoperatively with no great heterogeneity in the same muscle. At the thigh, adductors, sartorius and gracilis were less affected with respectively, 51%, 56% and 57% of fat fraction before surgery. Adductors and gracilis remained less affected after surgery. Increase in fat fraction was significant for sartorius and multifidus muscle ($p<0.05$). No significant difference was found for the other muscles studied.

Compared to previously reported results in the literature, fat infiltration was on average six times more than typically developing subjects [20].

Quality of life

None of the patients had any postoperative back pain; they were satisfied with the surgery. Patients and caregivers noticed an improvement in their daily life transfers.

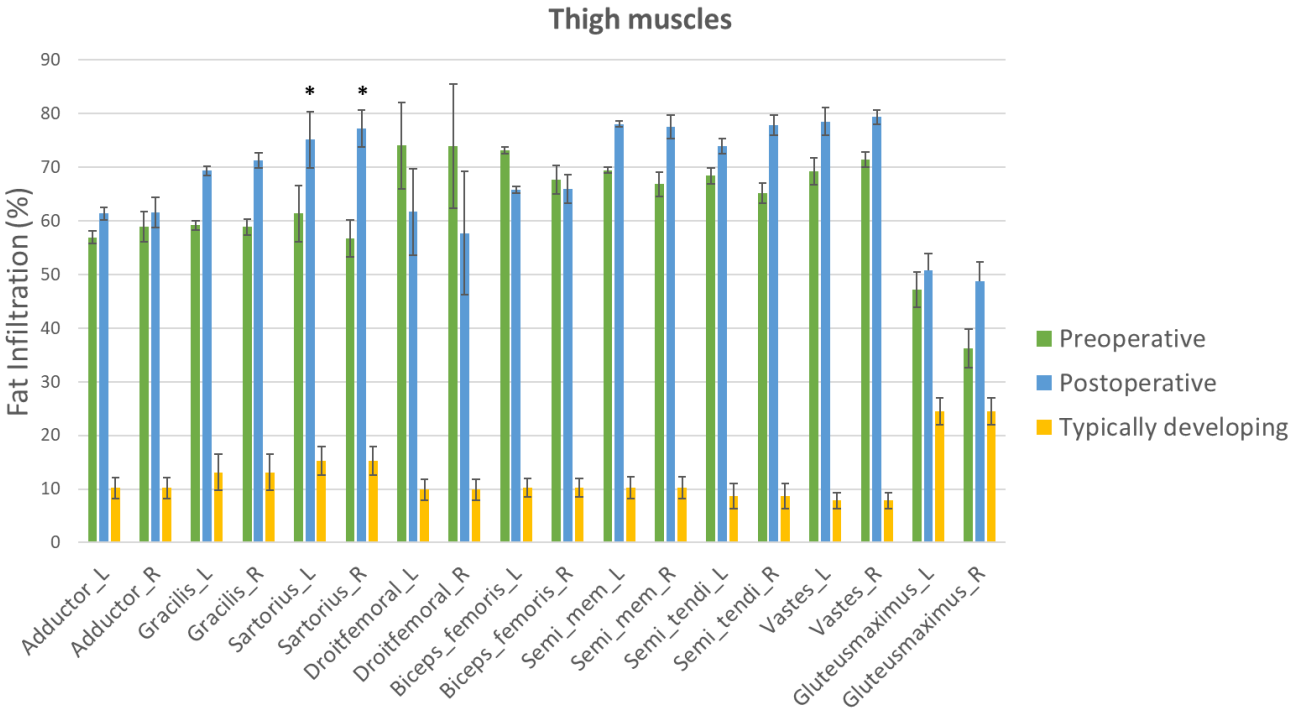

Figure 1: Fat infiltration at thigh level after and before surgery, and comparison with control subjects previously reported in the literature.

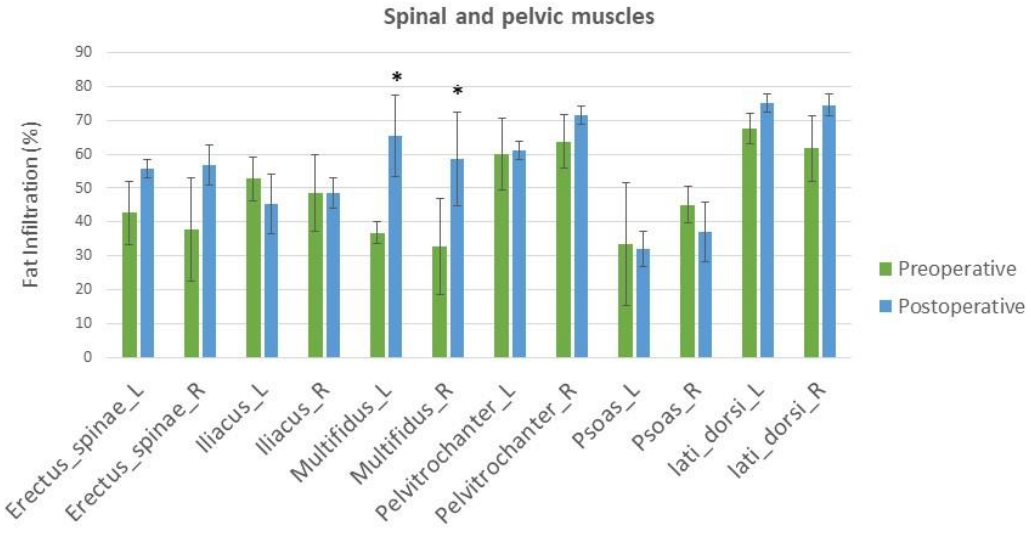

Figure 2: Comparison of fat infiltration at the spinal level, before and after surgery.

DISCUSSION

In this study, MRI of spinal and lower extremity muscles revealed diffuse and symmetric intra-muscular fat infiltration in both groups of SMA2 patients, which is characteristic of neuromuscular pathology. In contrast, a large intermuscular and interindividual variability of fat infiltration was previously observed with normal adults [21]. The sartorius, adductors and gracilis appeared less affected at thigh level, which is specific to SMA2 patients, in accordance to results previously reported [22-25].

Even if progressive nature of SMA over time is reflected by more severe fatty deposition after long disease duration [26-29], fatty degeneration in SMA remains slow with a yearly progression rate of less than 5% [30]. Consequently, the main differences observed at 5 years follow-up is mainly a consequence of the surgery. Many studies have reported the effects of posterior lumbar fusion surgery in adults on the paraspinal muscles by quantitative or qualitative analysis of MRI [31]. Sihvonen et al. pointed out that the cause of postoperative muscle atrophy was iatrogenic denervation of the paraspinal muscles [32]. The volume changes might be also the result of immobilization [33]. Some authors have observed decreases in muscle volume and increased fat infiltration of the spinal erector muscles in patients with a loss of lumbar lordosis [34-35]. Other studies have shown an association between increased fatty infiltration and low back pain [36-41]. Hyun et al. in a series of 44 operated adult scoliosis patients found that more than 60% of fat infiltration of the spinal erectors was a risk factor for postoperative junctional kyphosis [42].

In the present study, we were unfortunately unable to evaluate muscle atrophy because of artefacts related to respiratory movements or equipment. Unlike other studies [20], muscle volumes and inter-muscular infiltration were not analyzed due to the massive intra-muscular infiltration (figure 3).

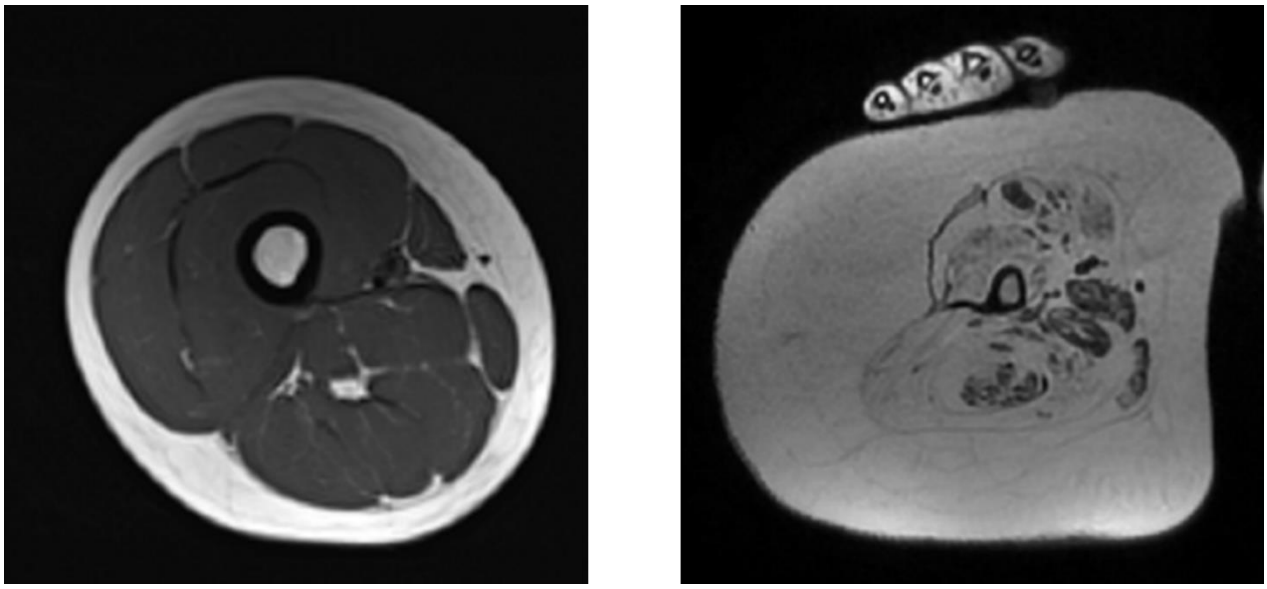

Figure 3. MRI muscle comparison at thigh between A/Control subject female; age 11 years, And B/SMA 2 preoperative female; age 10.5 years.

The increase in multifidus fat infiltration may be associated to the stability of the bipolar construct that prevented spinal mobility. However, the minimally invasive spinal surgery permitted to reduce muscle injuries, especially in the area between the proximal and the distal incision, which explains why the fat infiltration in erector spinae remained stable. Moreover, in the present study, a paraspinal (Wiltse) approach [43] was performed whereas the traditional distal midline posterior lumbar surgery is known to be a source of bilateral atrophy and fatty degeneration of the paraspinal muscles [34,44] (Figure 4). Thus, patients were comfortable in sitting position, with no trunk imbalance and no reported pain. Patients had probably more difficulties crossing their legs due to the correction of pelvic obliquity, leading to a lower activity of the sartorius muscle.

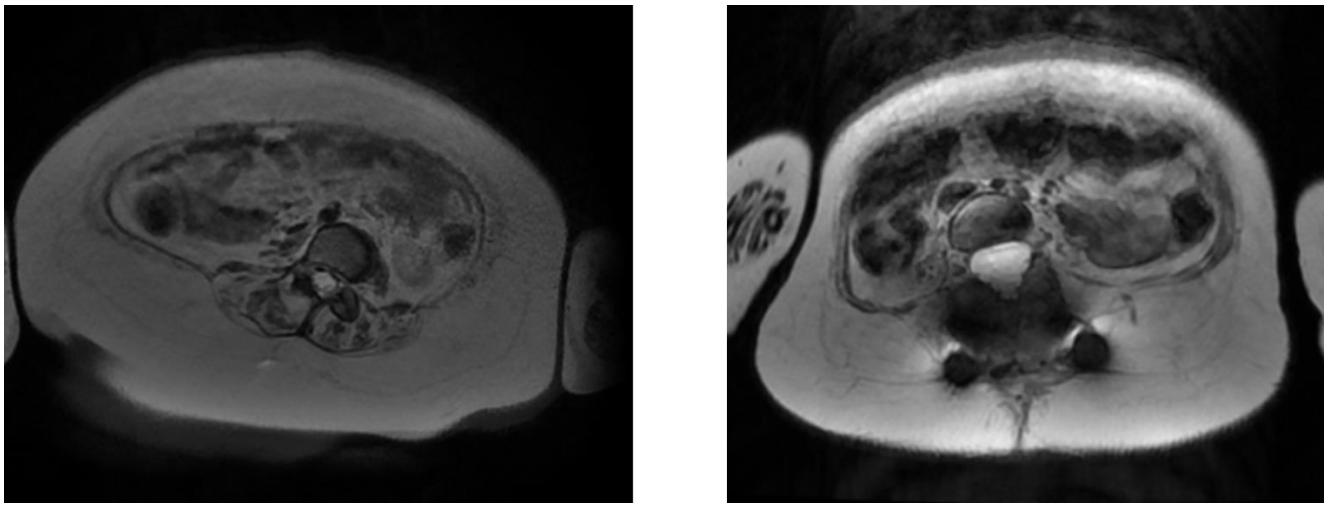

Figure 4. Paraspinal muscles MRI comparison A/Before surgery female; age 11 years, And B/After surgery preoperative female; age 17 years.

Our study has some limitations. Control subjects matching the patients could not be included, and the number of patients included was small, which may explain the absence of a significant difference in certain results. The small size of the cohort was explained by the low prevalence of SMA2 even in referral centers but was also exacerbated by the number of subjects who withdrew from the study because of the time-consuming nature of the acquisitions, which represented a significant additional burden for patients who already have frequent hospital visits, and thus were affected by a lack of motivation. If this study added some insight to better understand postoperative outcomes in this population, we do not think it is reasonable to do so in a future larger study unless new acceleration methods for muscle MRI are developed.

Conclusion

The present study is the first to provide objective MRI measures of fat infiltration after minimally invasive surgery in SMA2 patients. Our results demonstrate that muscles were globally preserved from fat infiltration after surgery apart from multifidus and sartorius that were more affected.

Patients	1	2	3	4	5	Mean
Age at MRI, years	18.0	19.0	20.0	17.2	16.3	18.1
Age at initial surgery, years	9.8	13.1	13.3	10.6	11.2	11.6
Mean follow-up, years	8.1	5.9	6.7	6.7	5.0	6.5
Halo gravity preparation	No	No	No	No	Yes	
Type of curve	TL right	Double major	Thoracic left	TL right	TL left	
Cobb angle of major curve,°						
Preoperative	103	92	60	86	102	88
Postoperative	40	48	30	21	62	40
Pelvic obliquity,°						
Preoperative	46	5	31	54	35	34
Postoperative	13	0	0	3	10	5
Kyphosis,°						
Preoperative	40	4	68	45	108	53
Postoperative	24	23	37	26	34	28
Lordosis,°						
Preoperative	47	15	83	52	56	50
Postoperative	43	21	25	37	51	36
Number of rod lengthening	2	1	1	0	0	
Vital Capacity (%)						
Preoperative	20	Nc	20	Nc	22	
Latest follow-up	23	Nc	32	Nc	26	
Complications (Mechanical/Infections)	0	0	0	0	0	0

Table 1: Main characteristics of postoperative patients . TL = thoraco-lumbar

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9. Conclusion générale et perspectives

L'objectif de cette thèse était d'étudier les modifications biomécaniques à long-terme induites par l'instrumentation sans greffe bipolaire chez les patients atteints de scoliose neuromusculaire. L'évolution clinique et radiologique de ces patients à plus de 5 ans de recul était stable, et nous avons étudié leurs particularités anatomiques sur le plan osseux, discal et musculaire.

Cette thèse a permis d'apporter des preuves objectives de l'enraidissement rachidien au décours de la chirurgie, et de proposer des outils et des méthodes pour le quantifier. Il s'agit d'une avancée significative pour permettre de justifier l'absence de nécessité d'arthrodèse finale lorsque les patients arrivent à maturité squelettique.

En pratique clinique, au regard de ces travaux, nous pourrions proposer pour les patients à maturité squelettique, la mise en place d'une élastographie discale lombaire, qui est un geste simple, rapide, non invasif et non irradiant de suivi. En cas d'enraidissement discal, celle-ci doit être complétée par un examen de tomodensitométrie avec un protocole à basse irradiation, afin de quantifier l'autofusion osseuse spontanée. L'IRM musculaire a permis de caractériser l'infiltration graisseuse de ces patients en pré et post opératoire. Cependant, cet examen n'est pas adapté pour justifier de l'absence de nécessité d'arthrodèse en fin de croissance, et nous ne le retenons pas pour le suivi.

L'instrumentation sans greffe bipolaire apparait donc une chirurgie peu invasive, avec un résultat stable dans le temps, et surtout définitive, tout en préservant la croissance thoracique et vertébrale. Elle permet une autofusion progressive, qui évite les risques liés à la conversion en arthrodèse. Les travaux réalisés nous permettent d'avoir un discours rassurant quant au spectre de la conversion du montage à maturité squelettique.

Cette thèse ouvre des perspectives de recherches, car nous sommes encore loin d'avoir élucidé tous les facteurs physiopathologiques qui interviennent dans cet enraidissement rachidien. Il serait intéressant de pouvoir l'accélérer ou le ralentir en fonction du potentiel de croissance restant. Il faudrait également parvenir à établir un arbre décisionnel permettant de définir l'absence de nécessité d'arthrodèse vertébrale postérieure, en fonction des arguments cliniques et d'imageries, réalisé à partir de cohortes multicentriques.

10. Références

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11. Liste des publications et communications liées à la thèse

11.1. Publications

-Gaume M, Vergari C, Khouri N, Skalli W, Glorion C, Miladi L.Minimally Invasive Surgery for Neuromuscular Scoliosis: Results and Complications at a Minimal Follow-up of 5 Years. ***Spine.*** 2021 Dec 15 ;46(24):1696-1704. doi: 10.1097/BRS.0000000000004082.

- Gaume M, Saudeau E, Gomez-Garcia de la Banda M, Azzi-Salameh V, Mbieleu B, Verollet D, Benezit A, Bergounioux J, Essid A, Doehring I, Dabaj I, Desguerre I, Barnerias C, Topouchian V, Glorion C, Quijano-Roy S, Miladi L. Minimally Invasive Fusionless Surgery for Scoliosis in Spinal Muscular Atrophy : Long-term Follow-up Results in a Series of 59 Patients. ***J Pediatr Orthop***. 2021 Oct 1;41(9):549-558. doi: 10.1097/BPO.0000000000001897

- M Gaume, T Langlais, K Loiselet, S Pannier, W Skalli, C Vergari, L Miladi. Spontaneous induced bone fusion in minimally invasive fusionless bipolar fixation for neuromuscular scoliosis: A computed tomography analysis. ***European Spine Journal,*** avril 2023

- M Gaume, K Loiselet, HChekir, T Langlais, N Boddaert, S Pannier, W Skalli, L Miladi, C Vergari. Evidence of Spinal Stiffening following Fusionless Bipolar Fixation for Neuromuscular Scoliosis. A Shear Wave Elastography Assessment of Lumbar Annulus Fibrosus. **(soumis*)***

- M. Gaume, C Vergari, M Creze, A Bonnet-Lebrun, C Muth-Seng, E Ferrero, S Pannier, S Quijano-Roy, L Miladi, W Skalli, R Carlier. Thigh and paraspinal muscles changes after fusionless bipolar fixation for early onset scoliosis in type 2 spinal muscular atrophy (**soumis)**

11.2. Communications

-16th Annual International Congress on Early Onset Scoliosis and the Growing Spine, Rome, 2022. Oral presentation : “Can We Definitely Avoid Spinal Fusion Surgery for Progressive Early Onset Neuromuscular Scoliosis.”

-41st European Paediatric Orthopedic Society, Krakow, 2023. Oral presentation : “Evidence of Spinal Stiffening following Fusionless Bipolar Fixation for Neuromuscular Scoliosis. A Shear Wave Elastography Assessment of Lumbar Annulus Fibrosus.”