Is Growth-Friendly Surgery Effective for the Treatment of Spinal Deformity in Patients with Arthrogryposis Multiplex Congenita?

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Introduction

Arthrogryposis Multiplex Congenita (AMC)

2+ congenital contractures

>150-300 disorders

Scoliosis: 30-70%

Effect rigidity?





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Surgical treatment of scoliosis in a rare disease: arthrogryposis

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> PEDIATRIC S P I N E STUDY GROUP

Background

AMC vs. AIS

Less correction Increased complications, progression

Reoperations

29% risk / 4Y Second highest (neurofibromatosis)

Escobar syndrome

Small subset AMC Outcomes: Escobar = IEOS





HARVARD MEDICAL SCHOOL TEACHING HOSPITAL J Neurosurg Pediatr. 2018 Oct 12;23(1):22-29. doi: 10.3171/2018.7.PEDS18347.

Case-matched comparative analysis of spinal deformity correction in arthrogryposis multiplex congenita versus adolescent idiopathic scoliosis.

Xu L, Chen Z, Qiu Y, Chen X, Li S, Du C, Zhou Q, Sun X.

Does Reoperation Risk Vary for Different Types of Pediatric Scoliosis?

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Growth-Friendly Spine Surgery in Escobar Syndrome

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- 1) To evaluate the outcomes of growth-friendly surgery (GFI) in patients with AMC and spinal deformity
- 2) To compare these results to a matched cohort of idiopathic EOS (IEOS) patients to determine whether the presence of perceived spinal rigidity and extremity contractures influenced outcomes







Methods: Study Design

Retrospective cohort: 2 EOS databases

Inclusion:

Diagnosis AMC Early-onset scoliosis Growth-friendly surgery

Exclusion:

Neuromuscular etiology >10 years old at GFI <2Y follow-up Inadequate data





AMC-related disorders

Arthrogryposis Amyoplasia Escobar syndrome Freeman-Sheldon syndrome Pterygium syndrome



Methods: Statistical Analysis

Propensity Score Matching

- Age
- Sex
- Curve magnitude
- Construct type



Linear mixed modeling

Comparison of curve across groups







Results

	AMC (n=35)		IEOS (n=112)		
Variable	Mean	Range	Mean	Range	Р
Age at GFI (years)	6.2	2.1-9.9	6.4	1.9-10	0.65
Construct (n; %)					0.69
TGR	16	46%	55	49%	X
VEPTR	8	23%	31	28%	- 7
MCGR	10	29%	18	16%	0
Shilla	1	3%	8	7%	
GFI Duration (years)	5	2.1-11.2	5.7	2-11.9	0.18

Similar: age, constructs, GFI duration







Results: Presentation





Results: Growth-Friendly Surgery



Results: Growth-Friendly Surgery

	AMC (n=35)		IEOS (n=112)		
Major Cobb (°)	Mean	Range	Mean	Range	Р
Pre-Index	76	34-101	75	40-130	0.65
Post-Index	49	21-84	42	12-88	0.03
Absolute Change	27	1-51	33	0-68	0.02
% Correction	35%	1%-56%	44%	0%-82%	0.01
Last GFI / Pre-Fusion	55	22-109	49	9-105	0.12
Post-Fusion	50	19-83	39	12-80	0.07







Results: Adverse Events

Median Complications 2 AMC vs. 1 IEOS (p=0.09)

Median Revisions

2 AMC vs. 1 IEOS (p=0.88)

Limitations

Complication reporting Sagittal deformity / T1-S1 Intraoperative difficulty >> Currently analyzed







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Conclusion

IEOS > AMC

Initial correction

IEOS = AMC

Final curves Complications Revisions

GFI is effective

More aggressive management in AMC?





