



Escobar Syndrome (Multiple Pterygium Syndrome) and Thoracic Kyphoscoliosis

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INTRODUCTION

- Escobar syndrome (ES) is characterized by a web across every flexion crease in the extremities, most notably the popliteal space.
- The syndrome is associated with other structural anomalies:
 - Vertical talus,
 - Thoracic kyphoscoliosis,
 - Severe restrictive lung disease.

PURPOSE

- In our study we evaluated two brothers diagnosed with Multiple Pterygium Syndrome type Escobar.



METHOD

- Two brothers (age 14 and 18) were diagnosed with ES via genetic analysis.
- The younger brother was diagnosed with
 - kyphosis and progressive scoliosis,
 - high palate,
 - ptosis,
 - low-set ear,
 - arachnodactyly,
 - faciocranial dysmorphism,
 - mild deafness and joint contractures.
- The older brother had the same symptoms but wasn't scoliotic.

METHOD

- Both received operations for
 - inguinal hernia,
 - PEV correction,
 - dislocated hip in early childhood.
- Cobb method was used to measure spine deformity.
- The younger patient received posterior vertebral instrumentation between T3 - L1.

METHOD

- Two brothers diagnosed with Multiple Pterygium Syndrome type

Name	Age/ Gender	Spine Related Pathologies	Structural Anomalies	Preop AP/LAT Cobb	Final AP/LAT Cobb	Enstrumentation Level	MRI	Follow-up (y)	Received Surgery
AT	14/Male	Thorasic Scoliosis, Kyphosis	Joints contractures, Faciocranial dismorphism	78/70	40/50 (Correction 48.8%)	Posterior/T3-L1	L5 sacralization, Tethered cord, vertebral fusion	2.5 (postop)	Hip, PEV, Ptosis, Inguinal Hernia
BT	18/Male	Kyphosis	Hip dislocation, joint contractures, dismorphism	3/85	3/85	-	Syringomyelia, vertebral fusion	3	Hip, PEV, Inguinal Hernia

RESULT

- Simple posterior stabilization of the deformity was achieved in the younger brother.
- The younger patient had a 78° scoliotic curve and 70° thoracic kyphosis.



AT, 14 y, Preop

RESULT

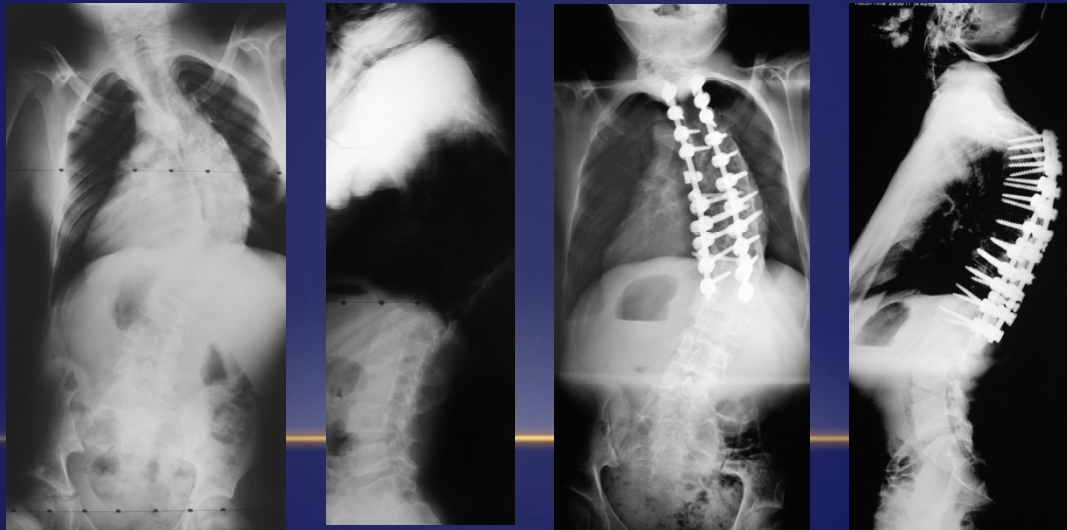
- Postoperatively correction rate: 48.8% (40°).



AT, Post-op

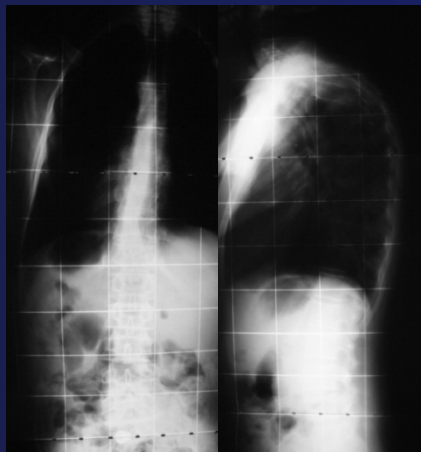
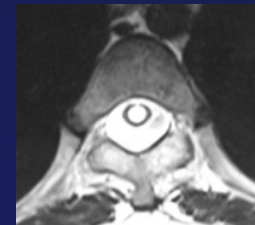
RESULT

- A follow-up period of 2.5 years revealed that the correction rate was maintained but the patient developed proximal kyphosis and a scoliotic curve progression in the lumbar area.
- For the younger patient MRI revealed
 - L5 was sacralised,
 - cauda equine level was at L3 and
 - proximal thoracic fusion



RESULT

- The older patient was diagnosed with 83° kyphosis;
 - syringomelia and
 - promixal thoracic fusion.



BT, 18 y

RESULT

- BT, 18 y



CONCLUSION

- Spinal pathologies commonly occur in patients with ES.
- Scoliosis may progress considerably over time.
- Early surgical treatment is recommended.
- However junctional kyphosis and/or progressive scoliosis in non-fusion areas can develop after corrective surgery.