

# Strategies to Improve Shoulder Range of Motion on a Patient with Loeys Dietz Syndrome s/p Subclavian Aneurysm and Pseudoaneurysm Repair. A Case Study.

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## Abstract

**Case Diagnosis:** Loeys-Dietz syndrome

**Case Description:** 54 y/o F diagnosed with Loeys-Dietz syndrome, admitted to inpatient rehabilitation facility (IRF) after 4 surgeries in the previous two months for repair of left subclavian aneurysm and pseudoaneurysm, left carotid to axillary bypass, left brachial stenting. Prior to her surgeries, she reported normal ROM of her left shoulder.

At IRF, goal was to maintain/increase the left shoulder ROM and to increase the level of independence with Activities of Daily Living (ADLs). On initial examination, patient was noted the following ROM: left shoulder flexion passive range of motion (PROM) 0-100 degrees, 0 degrees active range of motion (AROM), abduction not done to protect surgery sites. Upper extremity rehabilitation included tabletop slides, gentle shoulder AROM exercise, hand grip and dexterity exercises. Additional OT strategy/modification used for patient's shoulder rehabilitation was the use of a roller in a graded incline for ROM exercise.

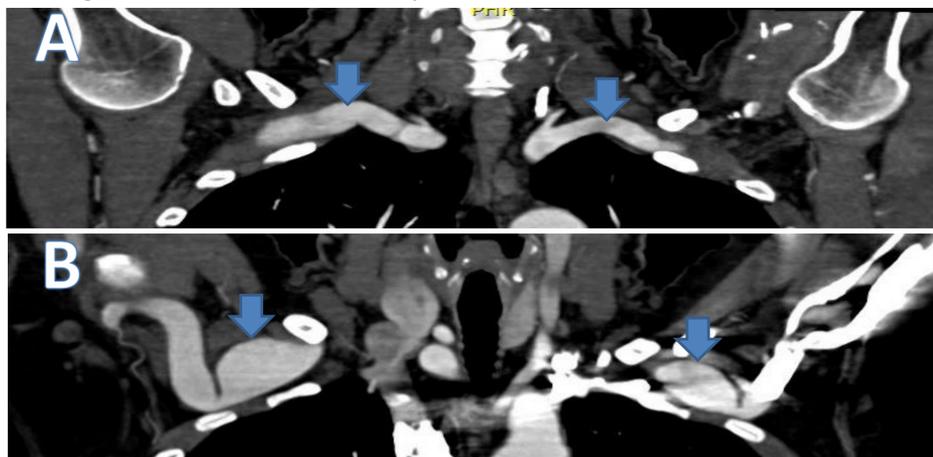
**Discussion:** At end of IRF stay, shoulder flexion was noted 0-100 deg PROM, 0-45 degrees AROM. Hand grasp measured by hand dynamometer increased from 0 lb to 1 lb. IRF stay totaled 13 days. Patient's ADLs improved to modified independent. UE dressing improved from moderate assist to modified independent. Patient continued with home therapies. After 6 months, muscle strength was 5/5 with 180° AROM and PROM. Functional independence measure (FIM) scores improved from moderate assistance to independent.

Loeys-Dietz syndrome is a rare autosomal dominant connective tissue disorder of the transforming growth factor- $\beta$  (TGF- $\beta$ )-related gene, characterized by aggressive arterial aneurysms. The only treatment is surgical repair of the aortic aneurysm to prolong life expectancy. Due to multiple surgeries of the left subclavian artery and axillary artery, the shoulder ROM and strength were affected. This case demonstrated importance of shoulder rehabilitation to achieve maximal ROM.

**Conclusions:** This case showed significant loss of upper extremity function in a patient with Loeys-Dietz syndrome following multiple surgeries. Modifying occupational therapy treatment and upper extremity exercises is important to improve shoulder functions and ADLs.

## Case Presentation

A 54 year old female with Loeys-Dietz syndrome was admitted to inpatient rehabilitation two months s/p multiple surgeries, including bilateral rib resections and left subclavian artery aneurysm and pseudoaneurysm repairs, left carotid to axillary bypass, left brachial artery stenting. Dilation of the patient's bilateral subclavian arteries can be seen in Fig. 1 (below). Prior to these procedures, range of motion and strength in her shoulders was reported to be within normal limits.



**Fig. 1 – Two slices from a single series of CTA imaging of patient's subclavian arteries prior to surgical bypass. Fig 1A demonstrates normal diameter of the subclavian artery medially. Fig 1B demonstrates dilation bilaterally as the arteries are followed laterally.**

After the surgeries, the patient had significant limitations to ROM and strength. On admission to IRF, the patient's PROM of left shoulder was 0-100° and she had 0° AROM. Upper extremity OT rehabilitation included typical treatments such as tabletop slides, gentle shoulder AROM, and hand grip and dexterity exercises. Additionally, the OT treatment was modified to include a graded incline roller seen in Fig. 2 (below).



**Fig 2 – Setup of graded incline roller. Patient seated in front of incline utilizes pectoralis, triceps, and contralateral deltoid activation to range the weakened shoulder through forward flexion. Angle of inclination could be increased as patient gained more strength in shoulder forward flexors. This therapy was well tolerated by the patient.**

## Results

	Admission to IRF	Discharge from IRF (13 day stay)	6 months s/p IRF
AROM in flexion (degrees)	0	45	180
PROM in flexion (degrees)	100	120	180
MMT (graded on 0-5/5 scale)	0/5	2/5	5/5
FIM for Upper Body Dressing	Min-Mod Assist (25-49% assist)	Modified Independent	Independent

## Discussion

A 54 year old female with Loeys-Dietz syndrome was treated at IRF s/p left subclavian artery aneurysm repair. Loeys-Dietz syndrome is a very rare autosomal dominant disorder in which the associated mutations in the TGFBR1 and TGFBR2 genes were discovered in 2005 study of 52 families. The prevalence of the disease is unknown. Unfortunately, the only treatment at this time is surgical repair. At the Cleveland Clinic, there were only 53 patients with Loeys-Dietz syndrome who had undergone medical or surgical treatment between 2005-2017. The graded incline roller was a novel therapy modality to accommodate for post-operative restrictions and the patient demonstrated improvements in function and ROM of the shoulder during and after IRF stay.

## Conclusions

Loeys-Dietz syndrome is a rare, Marfan-like syndrome that affects the vasculature leading to increased rates of arterial dissection, aneurysm, and pseudoaneurysm. In this case, the patient had marked loss of UE function s/p vascular surgical intervention to the subclavian artery. The patient demonstrated improvements in ADLs and function following aggressive IRF therapy regimen with modified occupational therapy for the shoulder.

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