

Surgical Outcomes in Juvenile Craniofacial Linear Scleroderma Patients: A 10-Year Single Center Cohort

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BACKGROUND

En coup de sabre, Parry-Romberg syndrome, mixed morphea, and circumscribed morphea are all subtypes seen in Juvenile Linear Scleroderma (JLS). JLS can cause marked facial asymmetry which can be corrected with various procedures:

- Autologous fat grafting
- Hyaluronic acid injection
- Implant
- Free tissue transfer
- Rhinoplasty
- Orthognathic surgery

PURPOSE

To describe long-term outcomes for pediatric patients who undergo surgical treatment for craniofacial JLS.

METHODS

- Retrospective review of 77 JLS patients from 2012 – 2023
- Data collected:
 - Disease onset, duration, and treatment history
 - Surgical details
 - Age at surgery
 - Intervention specific (cc fat injected)
 - Complications
 - Postoperative
 - Relapse
 - Skin quality
 - Need for additional procedures



Fig 1: 8-year-old with en coup de sabre affecting left facial nerve function with resultant asymmetric spontaneous smile.



Fig 2: 1.5 years following single-stage free innervated gracilis transfer to the masseteric branch of cranial nerve V.

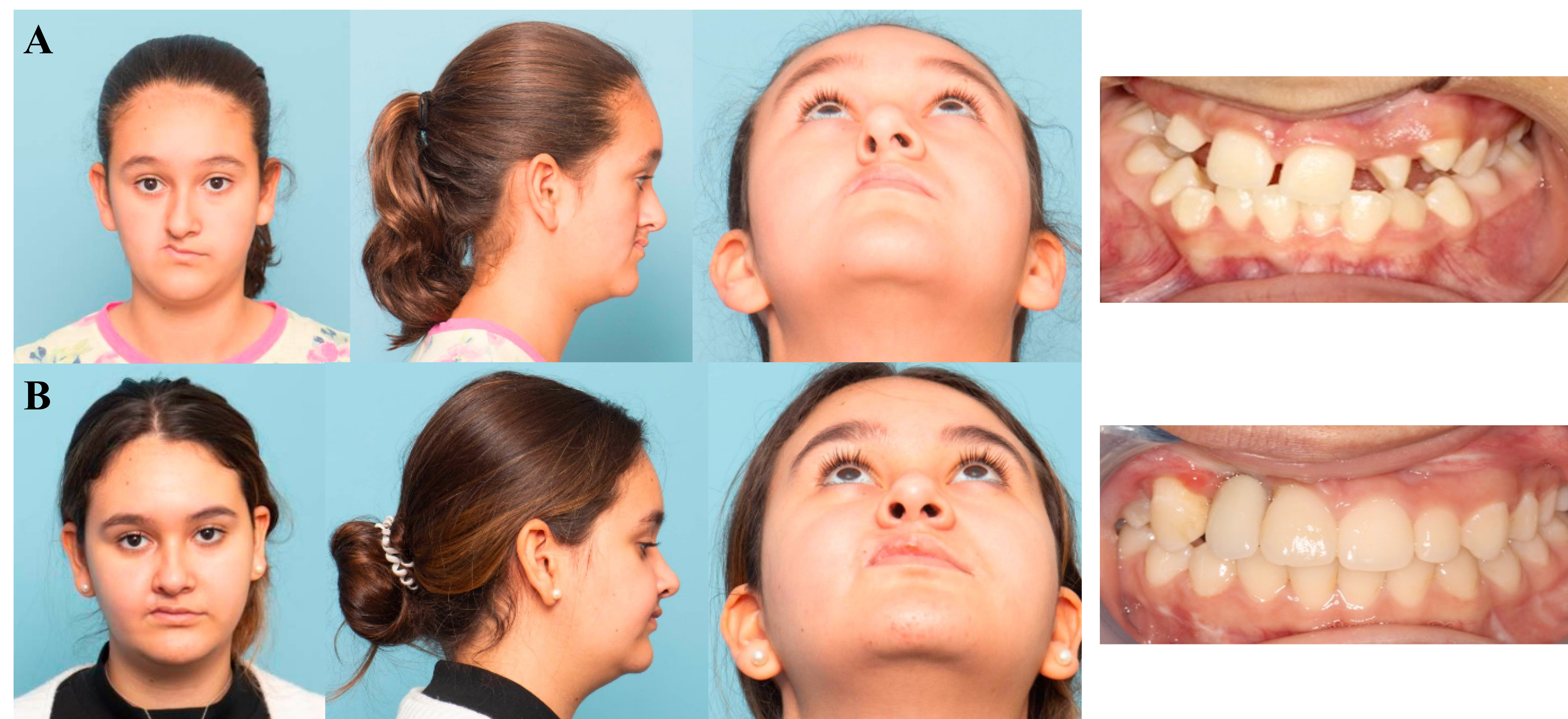


Fig 3: A) 13-year-old with Parry-Romberg syndrome **B)** Patient is shown 3 years later following 3 fat grafting sessions, orthognathic surgery, and septorhinoplasty.

RESULTS

- A total of 24 surgical patients were evaluated
- Patients had surgery at age 14.5 on average
 - Average duration of disease prior: 7.4 years
 - Years in remission prior: 4.8 years
- 1/3 of patients (n=7) were on disease-modifying medications at surgery
- Majority of patients required multiple procedures (2.4)
 - Autologous fat grafting (n=20)
 - Rhinoplasty (n=2)
 - Implant (n =2)
 - Free tissue transfer (n=2)
 - Orthognathic surgery (n=1)
 - Biomaterial filler (n=1)
 - Dermal graft (n=1)
- Autologous fat grafting outcomes
 - Average: 2.3 sessions
 - Average volume per session: 20.3 cc
 - No complications

CONCLUSION

There were no significant differences in complications for patients that were on disease modifying medications at the time of surgery. Additionally, there were no significant differences between patients whose quiescent intervals were > or < 1 year prior to surgery. Overall complications were low (4.2%). Most patients who had autologous fat grafting were able to achieve satisfactory facial symmetry after 2 fat grafting sessions.

REFERENCES

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