Attitudes and Trends in the Treatment of Morphea: A National Study

Nicole Strickland, B.S., Gopal Patel, M.D., Heidi Jacobe, M.D.

Department of Dermatology, University of Texas Southwestern Medical Center, Dallas Texas, 75390, USA.

INTRODUCTION

What is morphea?

- Morphea (localized scleroderma) is an inflammatory disorder of adults and children.
- It leads to cosmetic and functional impairment due to excess collagen deposition in the dermis and subcutaneous tissues.
- Morphea is classified into several clinical subtypes (plaque, linear, generalized, mixed) based on depth and location of lesions.
- Linear morphea is the most common subtype of morphea in children; plaque is common in adults.

What is the current state of morphea management in the United States?

- The attitudes of the specialists caring for these patients (dermatologists and rheumatologists) have not been evaluated in regards to evaluation and treatment.
- There is no uniform standard of care for morphea.
- **Our hypothesis is that approach to evaluation and treatment of morphea is guided by the specialty training of the physician rather than disease characteristics.**

METHODS & MATERIALS

- A web-based cross-sectional survey was administered to physicians randomly selected from the current American Academy of Dermatology, Society of Pediatric Dermatology, and the American College of Rheumatology Membership Directories.
- The survey had 22 questions in 3 sections:
  1) **General Information** (physician/ practice demographics)
  2) **Treatment of Morphea** (parameters guiding treatment choice, 1st and 2nd line treatment regimens for different morphea subtypes, treatment outcomes)
  3) **Management** (efficacy, adverse reactions, evaluation of therapeutic response, treatment duration, referrals)
- Statistical analyses, including frequency counts X^2 tests, and Fischer exact test (if there were < 5 responses in a cell) were calculated in GraphPad Prism.
- Statistical significance was set at $P < .05$.

RESULTS

Fig 1. Parameters guiding treatment choice in evaluation of morphea (all subtypes).

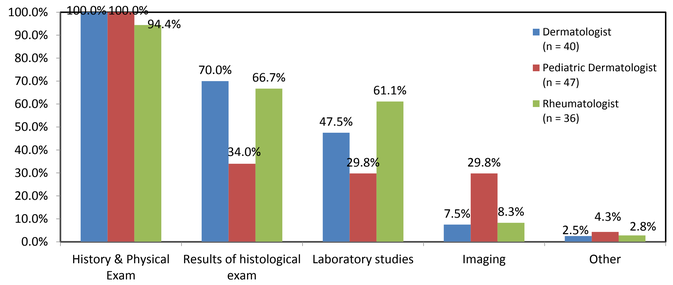

Fig 2a. Prescribed first-line treatments for plaque morphea.

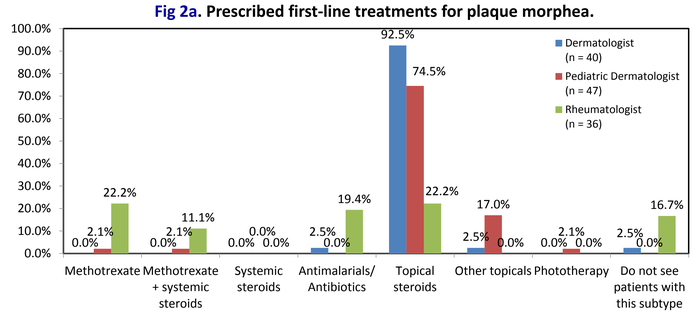

Fig 2b. Prescribed first-line treatments for linear morphea (not including en Coup de Sabre and Parry-Romberg subtypes).

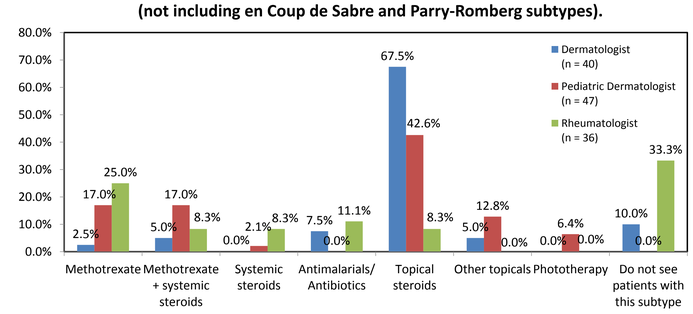

Fig 2c. Prescribed first-line treatments for en Coup de Sabre and Parry-Romberg subtypes of linear morphea.

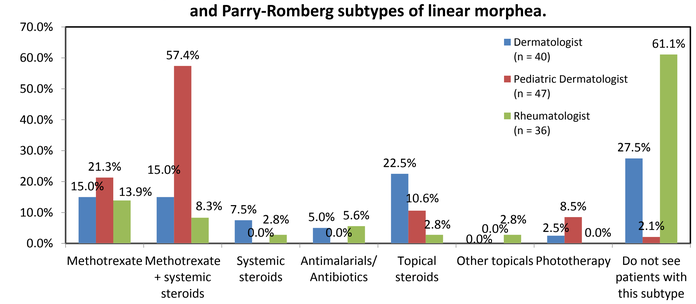

Fig 2d. Prescribed first-line treatments for linear morphea with lesions crossing a joint.

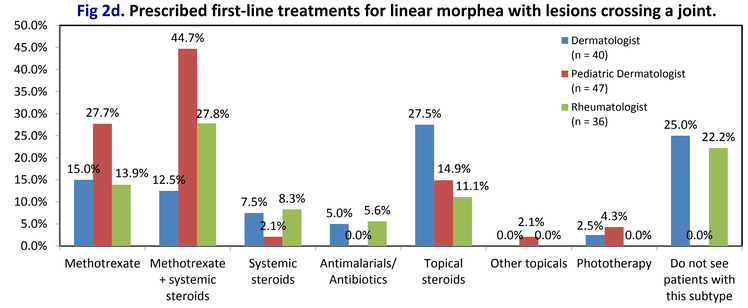

Fig 2e. Prescribed first-line treatments for generalized morphea.

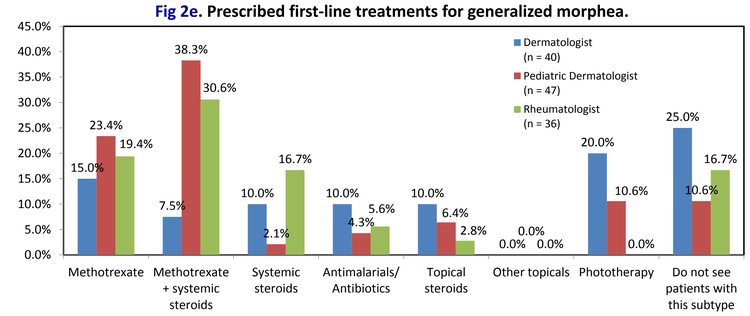

Table 1. Physician ratings of determinants of therapeutic response in morphea patients.

	Physician evaluation	Imaging (CT/ Ultrasound/ MRI)	Photography	Clinical Scoring System	Patient Perception
	1 = MOST important; 5 = LEAST important				
Dermatologist (n = 40)	1.1	4.49	3.58	4.2	1.82
Pediatric Dermatologist (n = 46)	1.04	4.02	2.7	4.27	2.74
Rheumatologist (n = 35)	1.09	4.62	3.97	3.54	1.82

Table 2. Duration of treatment for stable patients.

	Stop immediately	Continue < 1 month	Continue 1-6 months	Continue 6 months - 1 year	Continue > 1 year
	No. (%)				
Dermatologist (n = 40)	2 (5.0)	5 (12.5)	26 (65.0)	4 (10.0)	3 (7.5)
Pediatric Dermatologist (n = 46)	1 (2.2)	5 (10.9)	23 (50.0)	12 (26.1)	5 (10.9)
Rheumatologist (n = 35)	1 (2.9)	0	6 (17.1)	17 (48.6)	11 (31.4)

DISCUSSION

Evaluation of morphea

- The history and physical exam predominated decision making for all 3 specialties (Fig. 1).
- Pediatric dermatologists used imaging as an evaluation tool significantly more than the other specialties ($P = .0060$) and saw more linear morphea patients than their colleagues.

Treatment of morphea

- As shown in Fig. 2a-e, rheumatologists predominantly prescribed methotrexate ± systemic steroids and antimalarials/ antibiotics for plaque, linear, and generalized morphea.
- In contrast, dermatologists and pediatric dermatologists significantly favored topical steroids for plaque morphea, en Coup de Sabre/Parry-Romberg, and linear morphea lesions crossing a joint ($P < .0001$, $P < .0001$, $P = .0294$)
- Phototherapy was generally underutilized among all specialties except by dermatologists and pediatric dermatologists in treating generalized morphea ($P = .0181$).
- All 3 specialties ranked physician evaluation and patient perception as top determinants of therapeutic response (Table 1).
- Rheumatologists significantly continued therapy in stable patients for a longer period of time than other specialists ($P < .0001$) (Table 2).

Referrals

- The majority of dermatologists, pediatric dermatologists, and rheumatologists (73%, 83%, and 77%, respectively) referred morphea patients to their colleagues in other specialties.
- Dermatologists and pediatric dermatologists made referrals for functional limitations, possible or actual systemic findings, and for administering systemic therapy.
- Rheumatologists made referrals for confirming diagnosis and improving cosmetics.

CONCLUSIONS

- This study supports our hypothesis that physician training drives many aspects of morphea evaluation and treatment by dermatologists and rheumatologists.
- This study:

1) provides support for the necessity to develop evidence-based practice guidelines;
2) emphasizes the need for interdisciplinary collaboration that is already in place among these stakeholders in morphea care; and
3) shows a national perspective on morphea care that can be used as a platform to launch consensus statements on morphea treatment.

SELECTED REFERENCES

- Fett, N. and V.P. Werth, Update on morphea: part I. Epidemiology, clinical presentation, and pathogenesis. J Am Acad Dermatol, 2011. 64(2): p. 217-28; quiz 229-30.
- Fett, N. and V.P. Werth, Update on morphea: part II. Outcome measures and treatment. J Am Acad Dermatol, 2011. 64(2): p. 231-42; quiz 243-4.
- Johnson, W. and H. Jacobe, Morphea in adults and children cohort II: Patients with morphea experience delay in diagnosis and large variation in treatment. J Am Acad Dermatol, 2012.
- Li, S.C., et al., Development of consensus treatment plans for juvenile localized scleroderma. Arthritis Care Res (Hoboken), 2012.
- Li, S.C., et al., Treatment of pediatric localized scleroderma: results of a survey of North American pediatric rheumatologists. J Rheumatol, 2010. 37(1): p. 175-81.
- Zwischenberger, B.A. and H.T. Jacobe, A systematic review of morphea treatments and therapeutic algorithm. J Am Acad Dermatol, 2011. 65(5): p. 925-41.