# Pilot use of 3-dimensional photography to aid clinical decision-making in craniofacial morphea



*Key words:* craniofacial morphea; en coup de sabre; frontoparietal linear morphea; hemifacial atrophy; localized scleroderma; Parry-Romberg syndrome; 3D imaging; Vectra.

## **INTRODUCTION**

Craniofacial morphea includes progressive hemifacial atrophy, also known as Parry-Romberg Syndrome, and frontoparietal linear morphea, or en coup de sabre. These conditions cause damage to skin, fat, muscle, cartilage, and in severe cases, bone.<sup>1</sup> Disease onset often occurs between 5 and 15 years, with early involvement indicating a worse skeletal prognosis and disfigurement.<sup>2</sup> Although the etiopathogenesis is not fully understood, craniofacial morphea is considered an inflammatory condition in which immunomodulatory medications can effectively halt progression.<sup>3</sup>

Three-dimensional (3D) photography is a technique for assessing facial contours, including evaluation of volume augmentation following reconstruction.<sup>4</sup> Mirror imaging can be used to compare one facial half with the other at a single point in time, highlighting gradations of asymmetry.<sup>5</sup> Tracking mirror-image heat maps over time can illustrate disease progression. Overlaying full facial 3D images from different time points is also possible.

Herein, we describe 2 pilot cases in which 3D imaging was used to more objectively confirm disease stabilization in patients with craniofacial morphea. To date, 3D imaging has not been reported for this purpose.

Funding sources: None.

Abbreviation used:

3D: 3-dimensional

## **CASE REPORT**

With institutional review board approval, facial images were captured and analyzed using VECTRA 3D Imaging System (Version 5.7.2, Canfield). 3D photographs were taken in the plastic surgery clinic by clinical and/or research assistants at sequential clinic visits, spaced typically once every 3 to 4 months. The facial midline was identified, and the surface topography was copied and reflected. Volume differences between the affected and unaffected sides were quantitatively depicted with heat maps illustrating the geography and severity of asymmetry.

### Case 1

A 9-year-old with a long-standing history of en coup de sabre presented for consideration of facial reconstruction. Initially diagnosed at age 5, when a hyperpigmented plaque on the left forehead rapidly developed atrophy, the child was treated with pulse intravenous methylprednisolone and

for their photographs and medical information to be published in print and online and with the understanding that this information may be publicly available.

Correspondence to: Ingrid M. Ganske, MD, MPA, Assistant Professor of Surgery, Harvard Medical School, Department of Plastic and Oral Surgery, Boston Children's Hospital, 300 Longwood Avenue, Boston, MA, 02115. E-mail: ingrid. ganske@childrens.harvard.edu.

JAAD Case Reports 2022;25:100-3.

2352-5126

https://doi.org/10.1016/j.jdcr.2022.05.038

From the Department of Plastic and Oral Surgery, Boston Children's Hospital, Boston, Massachusetts<sup>a</sup>; the Department of Dermatology, Brigham and Women's Hospital, Boston, Massachusetts<sup>b</sup>; the Department of Dermatology, University of California Irvine School of Medicine, Irvine, California<sup>c</sup>; the Department of Pediatrics, Rheumatology Division, University of Pittsburgh Medical Center Children's Hospital, Pittsburgh, Pennsylvania<sup>d</sup>; Rheumatology Program, Boston Children's Hospital, Boston, Massachusetts<sup>e</sup>; and Dermatology Program, Boston Children's Hospital, Boston, Massachusetts.<sup>f</sup>

IRB approval status: Reviewed and approved by Boston Children's Hospital Institutional Review Board (IRB-P00029401). Consent for the publication of all patient photographs and medical information was provided by the authors at the time of article submission to the journal stating that all patients gave consent

<sup>© 2022</sup> by the American Academy of Dermatology, Inc. Published by Elsevier, Inc. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/ 4.0/).



**Fig 1.** Case 1: Girl with a history of en coup de sabre since age 5. Frontal, three-quarters, and lateral views of 3D photographs are shown at 6 time points using mirror-image analysis (**A-F**) and time-point overlay (**G**). *Yellow* indicates minor and *red* indicates greater volume deficiency on the heat map side compared with the contralateral side. **A**, Vertical depressions noted in the paramedian and left central forehead, as well as slight asymmetry of the alar base, zygoma, and temple, were noticed at age 7 before resuming treatment. Follow-up imaging 6 (**B**), 12 (**C**), 18 (**D**), 21 (**E**), and 24 (**F**) months later demonstrating stable appearance. Questionable disease progression in the zygomatic region in **E** and **F** prompted additional time-point overlay from 21 to 24 months (**G**), which revealed no differences in this region (*green*). Note that inconsistent facial expression may make assessment of the lower face less reliable.

subcutaneous methotrexate. Two years after the disease was thought to have stabilized, the family sought consultation for facial restoration; however, the substantial interval of facial growth and development limited the direct comparison of previous 2-dimensional photographs. In preparation for reconstruction, serial 3D images were evaluated to confirm the stability of facial asymmetry (Fig 1).

## Case 2

A teenage patient with progressive hemifacial atrophy was imaged as the disease progressed. Six years prior, he noted left temple and scalp atrophy and alopecia, with similar changes on the right side a year later. He was diagnosed with bilateral progressive hemifacial atrophy and was treated with pulse intravenous methylprednisolone and subcutaneous methotrexate, which were later tapered and discontinued after disease stabilization. As a late teenager, he developed right temple disease progression and the medications were resumed. 3D images were evaluated using mirror-image analysis (Fig 2), revealing a decreased volume in the right temple compared with the left. Subsequent evaluations 3, 6, and 9 months following treatment reinitiation showed stable volumetric heat maps, corroborating disease stabilization.

#### DISCUSSION

Existing tools to track disease progression in craniofacial morphea are limited. Assessment traditionally relies on 2-dimensional photography, clinical notes, and provider/patient recall. Inventories such as the Localized Scleroderma Cutaneous Assessment Tool are helpful globally but not designed to detect subtle evolving facial atrophy.<sup>6</sup> Comparing 2-dimensional photographs from multiple time points is often confounded by inconsistent facial expression, interval growth, and weight changes.

3D photography can help assess patients with craniofacial morphea by volumetrically assessing facial contours. Comparing the mirrored face at sequential time points detects relative asymmetry while controlling for growth and maturity. In Case 1, 3D imaging confirmed disease stability amid ongoing pediatric development. In Case 2, it confirmed the stabilization of a fully grown adolescent's disease. In both cases, 3D imaging complemented the clinical assessment by detecting stabilization or progression, thereby helping tailor medical management and aiding decision-making regarding preparedness for facial reconstruction. Surgical intervention is typically deferred until the disease has been stable for at least 2 years, although timing considerations remain an area of ongoing investigation.



**Fig 2.** Case 2: Boy with bilateral hemifacial atrophy presenting with disease flare of the right temple. Frontal, three-quarters, and lateral views of 3D photographs shown at 4 time points using mirror-image analysis. *Yellow* indicates minor and *red* indicates greater volume deficiency on the heat map side compared with the contralateral side. At age 18, before resuming treatment, 3D imaging captured right temple volume deficiency compared with the left (**1A-1C**). Minor asymmetry of the lateral chin was also noted. Imaging 3 (**2A-2C**), 6 (**3A-3C**), and 9 (**4A-4C**) months following retreatment showed a nearly identical pattern of atrophy without evidence of disease progression.

While this study was performed using a stationary 3D camera, technological advances have enabled the development of portable 3D cameras, which may make the implementation of this imaging even easier to incorporate into dermatology and rheumatology clinic locations, where patients are often primarily treated. With either stationary or portable cameras, the imaging analyses can be conducted by a seasoned clinical or research assistant within 5-10 minutes and could be incorporated into immediate decisionmaking in the clinic.

This preliminary report on 3D photography demonstrates clinical usefulness in following patients with craniofacial morphea and warrants further investigation to help care for patients with this challenging orphan disease.

#### **Conflicts of interest**

Dr Dedeoglu receives consulting fees from Novartis and royalties from *UptoDate*. Dr Ganske serves as an

uncompensated member of the governing board of The Foundation for Faces of Children. Authors Cappitelli, Langa, Min, Torok, and Vleugels have no conflicts of interest to declare.

#### REFERENCES

- 1. Tollefson MM, Witman PM. En coup de sabre morphea and Parry-Romberg syndrome: a retrospective review of 54 patients. *J Am Acad Dermatol.* 2007;56:257-263. https://doi.org/10.1016/j. jaad.2006.10.959
- Grippaudo C, Deli R, Grippaudo FR, Di Cuia T, Paradisi M. Management of craniofacial development in the Parry-Romberg syndrome: report of two patients. *Cleft Palate Craniofac J.* 2004;41:95-104. https://doi.org/10.1597/ 02-066
- Sommer A, Gambichler T, Bacharach-Buhles M, von Rothenburg T, Altmeyer P, Kreuter A. Clinical and serological characteristics of progressive facial hemiatrophy: a case series of 12 patients. J Am Acad Dermatol. 2006;54:227-233. https: //doi.org/10.1016/j.jaad.2005.10.020
- Hwang HS, Hwang CH, Lee KH, Kang BC. Maxillofacial 3-dimensional image analysis for the diagnosis of facial asymmetry. Am J Orthod Dentofacial Orthop. 2006;130: 779-785. https://doi.org/10.1016/j.ajodo.2005.02.021

JAAD Case Reports Volume 25

- Yu Z, Mu X, Feng S, Han J, Chang T. Flip-registration procedure of three-dimensional laser surface scanning images on quantitative evaluation of facial asymmetries. *J Craniofac Surg.* 2009; 20:157-160. https://doi.org/10.1097/SCS.0b013e318191ce88
- 6. Kelsey CE, Torok KS. The Localized Scleroderma Cutaneous Assessment Tool: responsiveness to change in a pediatric clinical population. J Am Acad Dermatol. 2013;69:214-220. https://doi.org/10.1016/j.jaad.2013.02.007