

Psychosocial considerations in patients and families affected by Freeman-Burian syndrome and other non-intellectually impairing craniofacial malformation conditions

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- **Contributors:** An anesthesiologist experienced in the care of craniofacial patients and two psychiatrists provided peer-review of this systematic review. Families and patients also contributed their perspectives.



Introduction / Background



- **Facial deformities** [e.g., Freeman-Burian syndrome (FBS)] engender less favorable social interaction
- May **affect patients / families / others psychosocially.**
- **Systematic review** conducted exploring:
 - 1) indirect or direct influence of non-intellectually impairing craniofacial malformation conditions on the psychosocial functioning of patients and families
 - 2) functional outcomes (educational, occupational, quality of life, etc.) of that influence



Methods



- **Searched** PubMed on 26 Sept 2017 and 18 July 2018.
- **Considered** all published papers meeting the search criteria. FBS handled separately. Searches re-run at end.
- **No meta-analysis** was attempted.
- **Review activities** were independently conducted by 2 experienced and trained reviewers. A third reviewer audited the process. No formal deliberative procedure was used.
- **Update** of Carson and Dufresne (1992)
- Part of **clinical practice guideline development** for FBS



Results: General / FBS



- **All conditions except FBS:** 738 unique citations, with 156 papers meeting inclusion criteria.
- **FBS:** 724 unique citations, with 101 papers identified as describing FBS and 5 meeting the inclusion criteria.
- **Exclusions:** lack of relevance, availability, or inability to translate.
- **5 Articles:**
 - High academic achievement; successful employment
 - Affect ranging from depressive/anxiety symptoms to “extroverted”



Results: Facial Aesthetics



- **22 Articles:**

- Facial unattractiveness / deformity created emotional distress; impacted quality of life and functional outcomes; and among the least desirable handicaps
- Social actions and determinants—not deformity severity—more important for psychosocial outcomes
- Larger facial deformities predicted stigma
- Persons viewing photographs consistently rate the craniofacially deformed individuals worse, unless rater had craniofacial deformity experience



Results: Psychosocial Confounds



- **6 Articles:**

- Facial appearance, not dysphasia, determined healthy children's attitudes toward the craniofacially deformed
- Dysphasia may be more psychosocially relevant
- Lower and less positive social engagement among adolescents and children than those not craniofacially deformed
- Manner in which young people present themselves predicted social perception
- Social skills training may improve social interaction



Results: Other Conditions



- **46 Articles:**

- More mood symptoms and worse psychosocial functioning in the craniofacially deformed children
- Symptoms / psychopathologies may not be a universal concern
- Impacts continue into adulthood
- Incongruence between self-report and objective assessment for psychosocial adjustment in adolescents
- Psychiatric and psychosocial outcomes depended on multiple variables, not just facial deformities



Results: Parenting Perspectives



- **5 Articles:**

- Parents may report more psychosocial difficulties or worse quality-of-life for their children than patients themselves
- Mothers were pessimistic about their children's coping strategies and impact of parental support





Results: The Social Self / Gender



- **12 Articles:**

- Difficulties with self-image (not body image) and self-concept are common but not universal
- Better quality of life predicted by better self-concept, resiliency, and absence of depression
- Global self-worth may be affected

- **16 Articles:**

- More studies found a worse outcome for girls, but others suggest girls and boys may be affected similarly
- Worse psychiatric outcome for adult men



Results: Positives / Misinformation



- **5 Articles:**

- Patients viewed having a deformity positively
- Experience may be psycho-protective
- Many children develop helpful coping mechanisms
- Many are well-adjusted adults, who did not feel deformities controlled their lives

- **3 Articles:**

- Positive and negative studies on maternal satisfaction with care team communication exist
- Dissatisfied mothers may have poor coping strategies



Results: Attitudes of Others



- **13 Articles:**

- Some attribute deformities to supernatural causes
- Other cultures' parents see a defect, adapt slowly, and are anxious about dysphasia and visible scarring
- Mothers of infants were anxious, unable to be sensitive / interactive, denied negative effects on parenting
- Less attractive infants = secure attachment (1 study)
- Five studies - no difference in infant-mother interactions
- Parents more lenient and teachers less tolerant
- Adults may continue to experience discrimination



Results: Tragedy of the Birth



- **9 Articles:**

- Craniofacially deformed child's birth is similar to a death
- Up to 70% of parents are very shocked at their child's birth and show depressive symptoms 7 years later
- Mothers of craniofacially deformed children may have higher frequencies of depressive / anxiety symptoms
- Trauma of the birth and stress of the experience of a craniofacially deformed child extends to other adults with whom the child interacts, with each reporting stress reactions, horror and disgust, or improved outlook



Results: Psychosocial Burden



- **26 Articles:**

- Negative impact on the family generally
- Minority may have no (2 studies) or positive influence (7 study) on the family/ parents
- Parents generally had negative impacts (impacting child's development): coping mechanisms, having a younger child, prenatal diagnosis, amount / satisfaction with social supports, financial burdens, parental craniofacial deformity, and strained marital relationship
- Elevated divorce rates



Conclusion / References



- **Craniofacial deformities** had a negative impact on most individuals and families.
- **Other factors** (eg, emotional state of and bonding with the mother, social support, finding meaning in life, helping others, and faith) are also significant determinants in overcoming psychosocial challenges.
- **Future studies** with robust methodology are needed.
- **References / Slides:** <https://www.duplastics.com/research>
- **Contact:** research@duplastics.com

Identification and approaches for psychosocial considerations in patients and families affected by Freeman-Burian syndrome and other non-intellectually impairing craniofacial malformation conditions: principles for global treatment

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ABSTRACT

Background: Many patients with facial deformities, such as Freeman-Burian syndrome (FBS), are treated less favorably than healthy individuals, which may affect patients psychosocially. We conducted a systematic review to explore the (1) indirect or direct influence of non-intellectually impairing craniofacial malformation conditions on the psychosocial functioning of patients and families and (2) functional outcomes (educational, occupational, quality of life, etc.) of that influence.

Methods: In searching PubMed on 26 Sept 2017 and 18 July 2018, we considered all published papers meeting the search criteria. FBS was handled separately. Searches were re-run before final analysis. No meta-analysis was attempted; the results were descriptively written to be practically useful and structured around the type or timing of psychosocial problems or consequences described or target population characteristics. All review activities were independently conducted by 2 experienced and trained reviewers. A third reviewer audited the process. No formal deliberative procedure was used.

Results: For all conditions except FBS, 738 unique citations resulted in 156 papers meeting the inclusion criteria. For FBS, 724 unique citations resulted in 101 papers identified as describing FBS, with 5 meeting the inclusion criteria. Most exclusions were for lack of relevance, availability, or inability to translate. Most included papers found more mood symptoms, impaired psychosocial functioning, and worse functional outcomes in craniofacially deformed children and their parents than for healthy children. Some found impacts continue into adulthood. Most papers found that psychiatric and psychosocial outcomes depended on multiple variables and not exclusively on objective facial deformities.

Conclusion: Craniofacial deformities had a negative impact on most individuals and families, but evidence also suggests other factors (eg, emotional state of and bonding with the mother, social support, finding meaning in life, helping others, and faith) are significant determinants in overcoming psychosocial challenges. Future studies with robust methodology are needed to confirm these findings.

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BIOSKETCH

Mikaela I Poling is an unpaid Research Assistant in plastic surgery. She has nearly two decades of experience in craniofacial clinical research, is an Associate Editor of the Journal of Pediatric Genetics and reviewer for a number of journals (including The Cleft Palate-Craniofacial Journal), has presented at several conferences, published over twenty papers, and led multidisciplinary research projects, including systematic reviews, a meta-analysis, and a clinical guideline task force. She has experience serving in various administrative, leadership, and community-engagement roles. In the early 2000s, she began mentored field research in limnology, geohydrology, and craniofacial surgery. She earned her undergraduate degree in history, with a specialization in modern Western military medicine and surgery. During college and while developing a long-range independent research plan with mentors in general surgery and plastic and craniofacial surgery, she completed several medical school clerkships. In particular, she gained supervised clinical experience caring for plastic surgery patients with full-thickness skin loss and multiple comorbidities. After completing undergraduate studies, she pursued advanced training in a clinical and applied physiology research fellowship. In this mentored setting, she continued her earlier work, obtaining a broad knowledgebase of human subjects research management and completing a graduate-level thesis, most of which has been published or is in review. Since 2016, she has been working with academic craniofacial surgeon, Craig R Dufresne, on projects of mutual interest. Together, they have presented and published work on Freeman-Burian syndrome, free dermal fat autografting for complex craniofacial wounds, and other topics.