Living with Moebius syndrome

Jenkinson, E., Bogart, K., Hamlet, C. and Davies, L.

Abstract: Moebius syndrome is a congenital neurological disorder that impacts facial expression, communication and appearance. In this article, the authors will discuss the psychological and social impacts of living with this rare form of facial palsy. Existing research suggests that patients may face challenges in developing psychological wellbeing, positive body image and in communicating effectively with others. Therefore, recommendations for nursing practitioners in how to best support this patient group are discussed.

Moebius (also known as Möbius or Mobius) syndrome is a rare congenital neurological disorder characterised by impaired facial expression and eye abduction (moving beyond midline) (Verzijl et al, 2003). It results from an absence or underdevelopment of the sixth and seventh cranial nerves (Verzijl et al, 2003; Briegel, 2006). The cause is unknown—it might be genetic or environmental in origin or a combination of the two. One theory proposes an ischemic cause, for instance, vascular disruption during early development (Briegel, 2006). Moebius syndrome is not heritable, although there are similar conditions that result in hereditary congenital facial paralysis, though eye abduction is not affected in these (Verzijl et al, 2003). The incidence is estimated to be one to 20 per million births, and it occurs equally in women and men and across races (Verzijl et al, 2003).

Moebius syndrome represents a spectrum of associated symptoms and severities. Bilateral facial and eye abduction impairment is most common, while unilateral manifestations are more rare (Verzijl et al, 2003). There are a number of other comorbidities that may be associated with Moebius syndrome, for example, most people have at least one of the following: strabismus; limb anomalies, especially of the hands and feet, such as brachydactyly and talipes equinovarus; microagathia, dysarthria, feeding difficulties (especially early in life); and motor and coordination problems. Incomplete closure of the eyes and mouth can lead to significant dry eye and corneal erosion complications and dry mouth, caries and orthodontic issues. There is controversy about whether autism and intellectual disability occur at higher rates in this population. Generally, earlier small studies reported higher rates (Johansson et al, 2001; Bandim et al, 2003), but more recent, larger studies using appropriate validated measures have not found increased incidence (Verzijl et al, 2005; Briegel et al, 2009a, 2009b; Briegel et al, 2010).

Moebius syndrome may affect many important life domains, including communication, socialisation and, sometimes, dexterity and motor tasks. Furthermore, Moebius is a visible difference, often with salient facial and hand differences. There is no specific treatment for Moebius syndrome; however, multidisciplinary supportive care is helpful to manage symptoms. Relevant fields include neurology, otolaryngology, orthopedics, ophthalmology, facial plastic surgery, medical genetics, dentistry, psychology and speech, physical and occupational therapy. Optional surgical interventions include facial reanimation surgery and hand surgery. There are several facial reanimation techniques, such as gracilis muscle transfer to the masseteric nerve or cross-face nerve graft (Zuker et al, 2000). These may improve speech, eating, dental health and the ability to smile (Goldberg et al, 2003).

Patients with Moebius syndrome may elect to engage in aesthetic procedures related to or unrelated to Moebius, but these should not be viewed as necessary or corrective. This could range from treatments for surgical scars, facial fillers to enhance symmetry or improve tone. However it is important to be aware that for patients with Moebius Syndrome, outcomes may not meet expectations. Aesthetic practitioners can play an important role in managing these expectations both at the initial consultation and during treatment to minimize negative psychological outcomes.

The psychological and social challenges of living with Moebius syndrome

While many with Moebius syndrome are psychologically adjusted and lead fulfilled lives, some may experience psychosocial difficulties. Firstly, facial expression serves as way to convey emotional information during our social interactions, and the reduced effectiveness of this important social communication tool is particularly challenging. Our smile communicates an intention to interact and socialise, and the reduced ability to smile can lead to others interpreting those with Moebius syndrome as uninterested or hostile, resulting in reduced or challenging social interactions (Bogart et al, 2014).

Appearance plays a crucial role in our self-concept (Moss and Carr, 2004), and the lack of facial expression in those with Moebius syndrome may not only affect how they relate to others, but how they relate to themselves. In a study exploring adults' experiences of living with congenital facial palsy, including those with Moebius syndrome (Davies, 2019), the central role the face plays in constructing the 'self' was highlighted. The facial paralysis often disrupted the facial mirroring process with others during early childhood, resulting in emotion regulation difficulties and resulted in some participants feeling unknown and misunderstood by others. This pattern often repeated across the lifespan and resulted in these individuals viewing themselves as 'defective'. Those who experienced validating relationships with others later in childhood or beyond were able to construct a validated and positive sense of self. This was achieved by developing non-verbal shared communication systems with others, which resulted in the individual with facial palsy feeling recognised and accepted on an affective and embodied level.

Limited research has explored the prevalence of mental health problems in the facial palsy population, but has generally found high levels of anxiety and depression (Fu et al, 2011; Pouwels et al, 2016). However, it is important to note that Moebius syndrome is congenital, and individuals may be better psychologically adjusted compared to those who acquire facial palsy later in life, whose self-concept is disrupted. Indeed, Bogart and Matsumoto (2010) found no increased incidence of anxiety or depression or decreased subjective well-being in 37 people with Moebius compared to a matched control group of people without facial palsy. In the largest study of people with facial paralysis (*n* = 546), Bogart (2020) found that both congenital and acquired participants had higher levels of anxiety and depression compared to the general public. Stigma was higher than norm data collected from other neurological conditions. The acquired sample, which primality consisted of people with Bell's palsy and acoustic neuroma, had significantly higher depression and anxiety compared to the congenital sample, with was primarily composed of people with Moebius syndrome. A multiple regression found the largest predictor of anxiety and depression was stigma.

Research suggests that people with ‘visible differences’ (Rumsey and Harcourt, 2004) and appearance-altering conditions experience enacted stigma (Goffman, 1963), such as name-calling, unwanted staring and avoidance by others, which can lead to social isolation (Masnari et al, 2012). Indeed, Moebius syndrome (and other types of facial palsy) is not widely recognised or understood among the general public (Bogart and Tickle-Degnen, 2015). In their qualitative research with adults and adolescents living with Moebius syndrome, Bogart et al (2012) and Bogart (2015) found that individuals had difficulties with social interactions, not only because of their lack of facial expression and confidence, but due to a fear of being negatively evaluated and previous experiences of prejudice. However, while communicating through facial expression was challenging, participants reported compensating by using expressive behaviours including body language and their voice (e.g. intonation) to express emotion. In a behavioural study, people with Moebius used more compensatory expression than those who acquired facial paralysis, suggesting an adaptive advantage (Bogart et al, 2012). Compensatory expression is an effective strategy for improving first impressions of people with facial paralysis; an experiment showed that people who used more compensatory expression were viewed as happier than those who used less compensatory expression, regardless of facial paralysis severity (Bogart et al, 2014).

Enhancing psychological wellbeing

The role of the nurse practitioner is pivotal in supporting patients with Moebius syndrome to make positive psychological adaptations by providing emotional containment and validation, and helping build social communication skills and positive body image.

As Moebius syndrome is a rare disorder, it can often be difficult for children, adolescents and adults living with the condition to have the opportunity to discuss their often unique lived experiences with others who understand and to access the emotional support that they might need (Bogart et al, 2017). This can result in feelings of loneliness and isolation and have a negative impact upon the individual’s psychological wellbeing (Briegel, 2006). Practitioners could play a key role in providing a safe and containing space for the individual to begin to explore any concerns that they might have about their appearance, medical/surgical procedures or challenging experiences they may have encountered as a result of living with Moebius syndrome.

Practitioners can build this therapeutic space by offering what Rogers (1957) termed the three core therapeutic conditions that are necessary for facilitating personal growth and emotional healing, and these are: empathy (understanding and compassion), unconditional positive regard (acceptance and care) and congruence (genuineness/realness). This can be achieved through actively listening to (paying particular attention to) the thoughts, feelings and wishes of the patient and reflecting them back through the use of the patient’s words and the use of appropriate facial expression and body language. This enables the individual to feel heard, understood and accepted on an emotional and embodied level and to feel safe enough to explore their experiences in an open and authentic way (Rogers, 1957).

Signposting these individuals to specialist support agencies may also help support the individual to develop good psychological wellbeing through accessing social support and meeting others with facial palsies. The Moebius Syndrome Foundation, an international organisation developed for individuals with Moebius syndrome and their families, moderates an online Facebook community that offers regular video chat/support, as well as occasional webinars. The Facial Palsy UK charity has a website that offers information/advice and face-to-face meetings/support groups held in various locations across the UK. Similarly, the UK-based charity Changing Faces provides information, support and advice to those living with a congenital or acquired visible difference.

Building social communication skills

Nurse practitioners can also provide psychosocial support to their patient with Moebius syndrome by facilitating the development of social communication skills and recognising and validating communication. This involves providing psycho-education around how we can use different parts of the body (e.g. hands, arms, shaking/nodding head, tone of voice, etc) to gesture and to express emotions non-verbally. This can be achieved by using a cognitive behavioural approach, whereby the nurse supports their patient to practice these non-verbal skills through engaging in skills practice/role-play and by providing them with constructive feedback in response (Clarke et al, 2012; Michael, et al, 2015). This may include practicing gesturing when conversing with family, friends or shopkeepers, and conversational skills. Additionally, preparation of responses for awkward questions about their appearance, as well as staring and negative comments from others may help patients gain confidence in social communication. When interacting with people with Moebius syndrome, health care professionals including aesthetic practitioners should pay special attention to alternative communication channels, including the voice and body language to facilitate understanding in consultations (Bogart and Tickle-Degnen, 2015).

Evidence-based online support developed at the Centre for Appearance Research in Bristol, UK, for individuals with congenital or acquired visible differences can be found through the Young Persons Face-IT programme or the Face-IT programme (for adults), which offer a specialist online cognitive behavioural skills training interventions for these populations. These online interventions have been found to be effective in reducing appearance-related distress in adults and young people with a range of appearance-altering conditions (Bessell et al, 2012, Williamson et al 2019).

Living with Moebius syndrome can present challenges in developing psychological wellbeing, positive body image and in communicating effectively with others. It is recommended that healthcare professionals respond to these challenges in interactions with patients through active listening, signposting to support from existing charities and considering how challenges to social communication may be overcome by cognitive behavioural therapy and social skills training. This might include referral to existing online interventions designed for patients with a range of appearance-altering conditions or psychological support. Patients with Moebius syndrome may also seek a range of wide range of aesthetic procedures. Aesthetic practitioners can play an important role in ensuring expectations regarding anticipated outcomes are managed appropriately to minimise dissatisfaction with clinical outcomes.

Useful links

Changing Faces: https://www.changingfaces.org.uk/

EURORDIS: https://www.eurordis.org/

Face IT: https://www.faceitonline.org.uk/

YP Face IT: https://www.ypfaceit.co.uk/

Facial Palsy UK: https://www.facialpalsy.org.uk/

Facial Paralysis and Bell’s Palsy Foundation: http://facialparalysisfoundation.org/

Moebius Syndrome Foundation: https://moebiussyndrome.org/

National Organization for Rare Disorders: https://rarediseases.org

Rare Disease UK: https://www.raredisease.org.uk/

References

Bandim JM, Ventura LO, Miller MT, Almeida HC, Costa AES. Autism and Möbius sequence: an exploratory study of children in northeastern Brazil. Arquivos de neuro-psiquiatria. 2003; 61(2A):181–185. https://doi.org/10.1590/s0004-282x2003000200004

Bessell A, Brough V, Clarke A, Harcourt D, Moss TP, Rumsey N. Evaluation of the effectiveness of Face IT, a computer-based psychosocial intervention for disfigurement-related distress. *Psychology, health & medicine*. 2012;*17*(5):565–577. https://doi.org/10.1080/13548506.2011.647701

Bogart KR. Socioemotional functioning with facial paralysis: is there a congenital or acquired advantage? Health Psychology. 2020; 39(4):345–354 https://doi.org/10.1037/hea0000838

Bogart KR. ‘People are all about appearances’: a focus group of teenagers with Moebius Syndrome. *J Health Psychol. 2015; 20*(12):1579–1588. http://doi.org/10.1177/1359105313517277

Bogart KR, Frandrup E, Locke T et al. ‘Rare place where I feel normal’: perceptions of a social support conference among parents of and people with Moebius syndrome. *Res Dev Disabil*. 2017; *64*:143–151. https://doi.org/10.1016/j.ridd.2017.03.014

Bogart KR, Hemmesch AR. Benefits of support conferences for parents of and people with Moebius syndrome. *Stigma and Health*. 2016; *1*(2):109

Bogart KR, Matsumoto D. Living with Moebius Syndrome: Adjustment, social competence, and satisfaction with life. *Cleft Palate Craniofac J.* 2010; *47*(2):134–142. http://doi.org/10.1597/08-257_1

Bogart KR, Tickle-Degnen L. Looking beyond the face: A training to improve perceivers’ impressions of people with facial paralysis. Patient Educ Couns. 2016; 98:251–256. https://dx.doi.org/10.1016%2Fj.pec.2014.09.010

Bogart KR, Tickle-Degnen L, Ambady N. Compensatory expressive behavior for facial paralysis: adaptation to congenital or acquired disability. Rehabil Psychol. 2012; 57(1):43–51. https://dx.doi.org/10.1037%2Fa0026904

Bogart KR, Tickle-Degnen L, Ambady N. Communicating without the face: holistic perception of emotions of people with facial paralysis. *Basic Appl Sol Psychol. 2014;* 36(4):309–320. https://dx.doi.org/10.1080%2F01973533.2014.917973

Bogart KR, Tickle-Degnen L, Joffe MS. Social interaction experiences of adults with Moebius Syndrome: a focus group. *J Health Psychol*. 2012; *17*(8):1212–1222. http://doi.org/10.1177/1359105311432491

Briegel W. Neuropsychiatric findings of Möbius sequence–a review. Clinical genetics. 2006; 70(2):91–97. https://doi.org/10.1111/j.1399-0004.2006.00649.x

Briegel W, Schimek M, Kamp-Becker I. Moebius sequence and autism spectrum disorders—Less frequently associated than formerly thought. Res Dev Disabil. 2010; 31(6):1462–1466. https://doi.org/10.1016/j.ridd.2010.06.012

Briegel W, Schimek Martina, Kamp-Becker I, Hofmann C, Schwab K. Autism spectrum disorders in children and adolescents with Moebius sequence. Eur Child

Adolesc Psychiatry. 2009a; 18(8):515–519. https://doi.org/10.1007/s00787-009-0003-1

Briegel W, Schimek M., Knapp D, Holderbach R, Wenzel P, Knapp E-M. Cognitive evaluation in children and adolescents with Möbius sequence. Child Care Health Dev. 2009b; 35(5):650–655. https://doi.org/10.1111/j.1365-2214.2009.00943.x

Clarke A, Thompson A, Rumsey N, Jenkinson E, Newell RJ. *CBT for Appearance Anxiety*. 1st edn. Hoboken (NJ): Wiley-Blackwell; 2013

Davies L. *The construction of 'self' in individuals with congenital facial palsy; a grounded theory exploration* (Doctoral dissertation). 2019. https://uwe-repository.worktribe.com/output/1490905 (accessed 4 April 2020)

Fu L, Bundy C, Sadiq SA. Psychological distress in people with disfigurement from facial palsy. *Eye (Lond)*. 2011; *25*(10):1322-1326. https://doi.org/10.1038/eye.2011.158

Goffman, E (1963) Stigma: notes on the Management of Spoiled Identity. Harmondsworth, UK: Penguin.

Goldberg C, DeLorie R, Zuker RM, Manktelow RT. 2003. The effects of gracilis muscle transplantation on speech in children with Moebius syndrome. Journal of Craniofacial Surgery. 14(5):687.

Masnari, O., Landolt, M.A., Roessler, J., Weingaertner, S.K., Neuhaus, K., Meuli, M. and Schiestl, C., (2012). Self-and parent-perceived stigmatisation in children and adolescents with congenital or acquired facial differences. *Journal of plastic, reconstructive & aesthetic surgery*, *65*(12), 1664-1670.

Michael, J., Bogart, K. R., Tylén, K., Krueger, J., Bech, M., Østergaard, J. R., & Fusaroli, R. (2015). Training in compensatory strategies enhances rapport in interactions involving
people with Möbius Syndrome. Frontiers in Neurology, 6, 1-11. doi: 10.3389/fneur.2015.00213

Moss, T. and Carr, T., 2004. Understanding adjustment to disfigurement: the role of the self-concept. *Psychology & Health*, *19*(6), 737-748.

Pouwels, S., Beurskens, C.H., Kleiss, I.J. and Ingels, K.J., 2016. Assessing psychological distress in patients with facial paralysis using the Hospital Anxiety and Depression Scale. *Journal of Plastic, Reconstructive & Aesthetic Surgery*, *69*(8),1066-1071.

Rogers, C. (1957). "The necessary and sufficient conditions of therapeutic personality change"*. Journal of Consulting Psychology.* ***21*** *(2): 95–103.*

Rumsey, N., and Harcourt D, (2005). The Psychology of Appearance. Open University Press.

Williamson, H, Hamlet, C., White, P et al. A web-based self-help psychosocial intervention for adolescents distressed by appearance-affecting conditions and injuries (Young Persons' Face IT): feasibility study for a parallel randomized controlled trial. JMIR Mental Health. 2019; 6(11). https://doi.org/10.2196/14776.

Verzijl H, Van Es N, Berger HJC, Padberg GW, van Spaendonck KPM. 2005. Cognitive evaluation in adult patients with Möbius syndrome. Journal of neurology. 252(2):202–207.

Verzijl HTFM, van der Zwaag B, Cruysberg JRM, Padberg GW. 2003. Mobius syndrome redefined: a syndrome of rhombencephalic maldevelopment. Neurology. 61(3):327.

Zuker RM, Goldberg CS, Manktelow RT. 2000. Facial animation in children with Mobius syndrome after segmental gracilis muscle transplant. Plastic and reconstructive surgery. 106(1):1.