



Quality of life and adjustment in children and adolescents with Moebius syndrome: Evidence for specific impairments in social functioning



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ABSTRACT

Introduction: Moebius syndrome is a rare congenital disorder characterized by congenital facial paralysis and impairment of ocular abduction. The absence of facial expression in people with Moebius syndrome may impair social interaction. The present study aims at replicating prior findings on psychological adjustment in children and adolescents with Moebius syndrome and providing first data on quality of life.

Participants and methods: Parents ($n = 26$) and children and adolescents ($n = 14$) with Moebius-Syndrome completed the KINDL^R (Questionnaire for Measuring Health-Related Quality of Life in Children and Adolescents), the SDQ (Strengths and Difficulties Questionnaire), and a newly devised questionnaire on the global, social, and emotional impact of Moebius syndrome, including a scale that addressed what conjectures were made concerning the thoughts of other people about a child with Moebius syndrome.

Results: In comparison with normative data, quality of life was reduced for the subscale *Friends* (large effect for both parental ratings and self-report) and the Total Score of the KINDL^R. Parents reported elevated levels of peer problems on the respective subscale of the SDQ. In SDQ self-report data, cases classified as abnormal were observed on *Peer Problems* only. Moebius-specific scales showed adequate reliabilities, and were related, most notably for parent-report, to quality of life. 29% of children reported feelings of anger when being stared at, and wished that they were not affected by Moebius syndrome.

Conclusions: Although most children and adolescents in our sample did not show behavioral problems or reduced quality of life, our results indicate that their emotional and social development should be closely monitored. Maintaining satisfying peer relationships seems to be a special challenge for children and adolescents with Moebius syndrome.

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1. Introduction

Moebius syndrome or sequence is a rare congenital, nonprogressive disorder characterized by paralysis of cranial nerves VII (facial nerve; controlling facial expression) and VI (abducens nerve; controlling lateral eye movement). Prevalence has been estimated at 0.002% of births (Verzijl, van der Zwaag, Cruysberg, & Padberg, 2003). Moebius syndrome can be associated with a variety of clinical findings like impairment of other cranial nerves, thoracic or limb anomalies, dental

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problems, malformation of the tongue and cleft palate. An increased rate of comorbid autism spectrum disorders has been observed (Johansson, Gillberg, & Råstam, 2010), but divergent findings have been reported as well (Briegel, Schimek, & Kamp-Becker, 2010). Studies on cognitive development are inconclusive. Intellectual disability may be present in 10–15% of individuals with Moebius syndrome (Briegel, 2006; Briegel et al., 2009).

Moebius syndrome can pose significant challenges for the development of social interaction and psychological adjustment (Briegel, 2006; Bogart, Briegel, & Cole, 2014; Bogart & Matsumoto, 2010; Bogart, Tickle-Degnen, & Joffe, 2012). Moebius syndrome is an immediately visible condition: Most people with Moebius syndrome cannot show any form of facial expression like smiling, frowning, or raising eyebrows, their face appears mask-like. The face, however, is an important target of attention in interpersonal contact and a source of socially relevant information. Facial expression is essential for effective and smooth social communication and establishing rapport (Tickle-Degnen, 2006). In particular, the ability to smile is crucial for maintaining reciprocal interactions and promotes social contact (Krueger & Michael, 2012; VanSwearingen, Cohn, & Bajaj-Luthra, 1999). Facial mimicry – the spontaneous imitation of facial expressions – intensifies feelings of connectedness (Krueger & Michael, 2012).

As Moebius syndrome is a rare condition, its features are mostly unknown to the public and thus difficult to interpret. The lack of facial expression may be irritating or perceived as uninterested, bored or depressed, especially when people with Moebius syndrome are unknown to others. Other features of Moebius syndrome like drooling or dysarthric speech may further increase the risk of social rejection and lead to stigmatization (Bogart, 2014) or may be misinterpreted as intellectual disability. The lack of facial expression may be compensated by verbal and nonverbal behavior (Bogart, Tickle-Degnen, & Ambady, 2012), however, effortful and less automated expressive behavior may inhibit the flow of natural communication (cf. Cole & Spalding, 2009).

In childhood impaired oral motor functions may also interfere with children's participation in play activities like inflating balloons or blowing soap bubbles (Broussard & Borazjani, 2008). Adolescence is characterized by a strong desire of belonging to peer groups, and self-esteem and mental health are strongly affected by social interaction experiences with peers (Brown & Larson, 2009). Therefore, adolescence may be a period of heightened vulnerability for youth with Moebius syndrome.

The impact of facial differences on social behavior, psychological adjustment and quality of life has been studied for various conditions (cf. Klassen et al., 2012; Rumsey & Harcourt, 2004, 2007). Masnari, Schiestl, Weibel, Wuttke, and Landolt (2013) found that children and adolescents aged 8–17 attributed less favorable characteristics to and were less willing to interact with children with facial differences. Stigma experiences, like being teased about their facial expression, were frequently reported by adolescents with congenital and acquired facial differences (Masnari et al., 2012; Masnari, Schiestl, Weibel, et al., 2013; Strauss et al., 2007; Tiemens, Nicholas, & Forrest, 2013) and related to psychological adjustment and quality of life (Masnari, Schiestl, Rössler, et al., 2013). Topolski, Edwards, and Patrick (2005) found significantly lower overall quality of life in adolescents with facial differences. In an observational study children with visible craniofacial conditions initiated less contact, received fewer positive reactions, and engaged less actively in conversations than their peers (Kapp-Simon & McGuire, 1997). Eiserman (2001) exploring positive contributions of facial differences, pointed out, that research was characterized by focusing deficits, although a majority the individuals studied fell in the nonclinical range on most variables.

From a developmental psychopathology perspective, negative evaluations of one's appearance (Harter, 2006) and disturbances in social relations may contribute to the development of psychiatric disorders (Sroufe, Duggal, Weinfield, & Carlson, 2000). Satisfying social relations and social competence are considered as important protective factors (e.g. Criss, Pettit, Bates, Dodge, & Lapp, 2002), whereas social exclusion and peer rejection enlarge the risk of developing psychiatric disorders (Ladd & Troop-Gordon, 2003).

Few studies, however, have addressed the specific effects of Moebius syndrome on social functioning and psychopathology of children and adolescents. Briegel, Hofmann, and Schwab (2007) found elevated levels of social problems in parental ratings of child behavior using the Child Behavior Checklist 4–18 (CBCL/4–18; Achenbach, 1999). Social problems of clinical relevance were found for 3 out of 12 adolescents, but only for one out of 19 children aged 4–11. Nevertheless, reported mean values of the respective scale were higher for the younger age group. In another questionnaire study by Briegel (2012) with 17 German participants aged 9–15 scores on the scale *Peer Problems* of the Strengths and Difficulties Questionnaire (SDQ; Goodman, 1997) fell in the abnormal range nearly three times as frequently as in a normative sample and showed a strong positive correlation with age. Self-reported levels of depression and anxiety were either in the normal range or even significantly lower. For preschool children Briegel et al. (2007) did not find increased rates of behavior problems.

Bogart, Tickle-Degnen, and Joffe (2012), investigating social interaction experiences of adults with Moebius syndrome in a focus group study, concluded that having a visible facial difference was experienced as stigmatizing. Participants reported experiences of teasing when they were children, resulting in low self-esteem. In another focus group study adolescents reported bullying and social strain (Bogart, 2014). Adults with Moebius syndrome reported lower social competence than a matched control group, but there were no differences for anxiety, depression, and satisfaction with life (Bogart & Matsumoto, 2010). Meyerson (2001) pointed out that people with Moebius syndrome may well achieve professional and personal success, relying on family support, faith, humor, determination and other sources of resilience.

In summary, most children, adolescents, and adults with Moebius syndrome do not show general maladjustment, but a substantial minority report impairment in social functioning.

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