

Psychosocial adjustment of 20 patients with Treacher Collins syndrome before and after reconstructive surgery

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Summary—Eight males and 12 females (mean age, 12.2 years) with Treacher Collins syndrome were studied longitudinally before and after craniofacial reconstruction. The patients and the parents of the 16 patients who were children were interviewed by a psychosocial team (child psychiatrist, psychologist and social worker) 6 months before and 1, 2, 3 and 4 years after surgery. The patients' facial appearance (Hay's Rating Scale), intellectual ability (Wechsler scales), self-esteem (Piers-Harris Self-Concept Scale for Children) and adaptive functioning (DSM III) were measured. The findings indicate that while their intellectual ability was unchanged, their appearance, self-esteem and adaptive functioning improved, peaking 1 year postoperatively and levelling off at the 2- and 4-year postoperative assessments. The improvement in the patients' facial appearance seems to have a direct, positive influence, creating psychosocial and social benefits for them.

Mandibulofacial dysostosis was first described by A. Thomson 140 years ago (Thomson, 1846). It was later named after E. Treacher Collins (Collins, 1900), who noted the marked malar hypoplasia that accounts for the flattened cheek region. More recent studies have described the morphologic features of the syndrome in greater detail (Francheschetti, 1944; Klimen, 1979; Smith, 1982; Kolar *et al.*, 1985). The psychological and social effects of facial disfigurement are known to be highly negative (Baker and Smith, 1939; McGregor, 1979; Berscheid, 1980), which points to the importance of plastic and reconstructive surgery in diminishing these consequences (Lefebvre and Munro, 1978; Munro, 1980; Strauss, 1980; Lefebvre and Barclay, 1982).

Treacher Collins syndrome is one of the most severe and potentially debilitating congenital anomalies. It is often accompanied by significant hearing loss, which constitutes an added hurdle to the patient's adaptation. However, systematic longitudinal studies involving patients with this syndrome, who have undergone facial reconstruction, are lacking. This report is the first from a prospective, longitudinal study measuring physical appearance and psychosocial adjustment of 20 Treacher Collins syndrome patients before and after facial reconstructive surgery performed at The Hospital for Sick Children, Toronto.

Materials and methods

Twenty patients with Treacher Collins syndrome (12 females and 8 males, aged 4.0 to 34.2 years, mean 12.2 years) and both parents of the 16 patients under 18 years old were interviewed by a psychosocial team during routine assessment. The team consisted of a psychiatrist, a psychologist and a social worker.

Preoperatively, demographic and family background information was collected. Patients and parents were also asked to explain their reasons for seeking facial reconstruction and were encouraged to discuss their physical and psychosocial expectations of it.

Each patient was rated on the DSM III, a classification measuring adaptive functioning, which ranges from 1 (superior) to 7 (grossly impaired) (American Psychiatric Association, 1980). The patients' full-scale intelligence quotient scores, comprised of verbal and performance measures, were also obtained, using age-appropriate Wechsler scales (Wechsler, 1974). The revised WISC has been defined and organised as a test of general intelligence. Its author believes that general intelligence exists; that it is possible to measure it objectively and that, by doing so, one can obtain a meaningful and useful index of a subject's mental capacity. The WISC-R has established itself as a

useful clinical and diagnostic tool. In addition, patients aged 6.0 to 14.0 years ($n=9$) were asked to complete the Piers-Harris Self-Concept Scale for Children, an 80 item yes-and-no questionnaire designed for children aged 6 to 18 years (Piers, 1969). The patients and parents were also asked to rate the preoperative facial appearance on the Hay's Rating Scale, which ranges from 1 (perfect feature) to 9 (marked imperfection) (Hay and Heather, 1973).

All patients underwent periorbital reconstruction and nine also had bilateral mandibular reconstruction.

At the time of the evaluations 1, 2, and 4 years postoperatively, patients and parents were again interviewed and all measures were repeated. The patients and their parents were also asked about the surgical experience, their satisfaction with the results and possible changes in the patient's life at home and school or work.

Differences between means for various groups and at different assessments were investigated with *t*-tests and an analysis of variance. The association between variables was measured with the Pearson correlation coefficient. Differences were taken to be significant when $p < 0.01$. The computer program package BMDP was used for all data processing and statistical analysis.

Results

Fifteen patients were Caucasian and five were of mixed racial background (black and white). Two patients came from Toronto, 13 from other areas of Ontario, one each from Nova Scotia and British Columbia, two from the United States and one from South America. At the time of the preoperative assessment, three-quarters of the parents were married and four couples were divorced or separated. No marital breakdown was noted during the study. English was the primary language spoken in 16 of the homes, Italian in two cases, Portuguese in one and Spanish in one. The mean number of children in the families was 3.05, of which 37.7% were normal and 62.3% had Treacher Collins syndrome. Four of the patients in the study had an affected parent and sibling, 8 had an affected parent or sibling and 8 had neither.

The clinical interviews revealed that all the patients over 6 years old and all the parents believed that facial surgery would facilitate integration into the normal world. The younger children were largely unaware of their facial anomaly. Fourteen

had mild to moderate hearing impairment, 3 had no hearing loss and the other 3 were severely hearing-impaired.

Almost two-thirds of the mothers were homemakers and all of the fathers were employed outside the home.

Mean adaptive functioning of the child as measured on the DSM III scale was just above the "poor" level preoperatively, peaked at "good" 1 year after surgery and settled just above "fair" 2 and 4 years postoperatively (Fig. 1). Repeated measure tests between the four variables showed a high F-ratio ($f=29.11$). All differences between preoperative and any postoperative ratings were highly significant (Table 1). The preoperative score correlated only with the 1-year postoperative score ($p=0.02$).

Table 2 illustrates the pre- and postoperative verbal, performance and full-scale intelligence scores in the Wechsler scales. All mean scores remained constant and in the normal range throughout the study period. Mean performance scores were significantly better than mean verbal scores at all four assessments (Table 3).

On the Piers-Harris Self-Concept Scale, the mean preoperative score was low, while the mean 1-year postoperative score was in the high range. Two and 4 years postoperatively the scores levelled off in the average range (Fig. 2). Repeated measure tests between the 4 variables showed a high F-ratio ($f=$

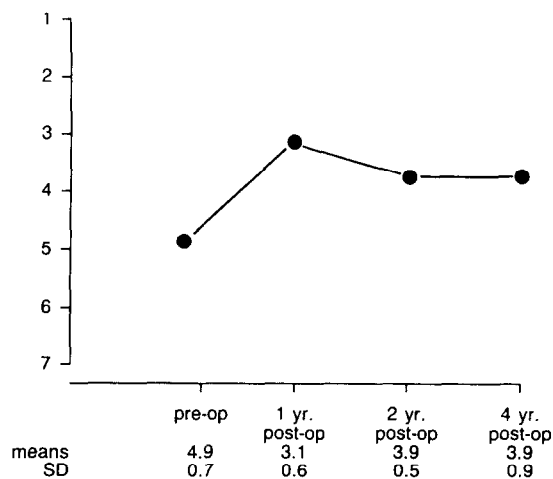


Fig. 1

Figure 1—Pre- and postoperative mean scores and standard deviations (SD) on the DSM III scale for 20 Treacher Collins syndrome patients. 5 = poor, 4 = fair, 3 = good

Table 1 Differences between and correlations of pre- and postoperative ratings for 20 Treacher Collins syndrome patients on the DSM III scale

Assessment compared with preoperative rating	Difference		Significance (p value)	Pearson correlation and (p value)
	Mean	SP		
1 year postoperative	1.9	0.6708	<0.0001	0.5181 (0.02)
2 years postoperative	1.0	0.8584	<0.0001	-0.0328 (NS)
4 years postoperative	1.0	0.1234	<0.0008	-0.0172 (NS)

Table 2 Pre- and postoperative mean scores and SD of 20 Treacher Collins syndrome patients on the age-appropriate Wechsler scales

Description of score	Assessment							
	Preoperative		1 yr postoperative		2 yr postoperative		4 yr postoperative	
	Mean	SD	Mean	SD	Mean	SD	Mean	SD
Verbal	93.6	13.5	93.05	12.8	93.7	10.7	92.1	10.7
Performance	103.2	12.1	106.2	10.8	106.8	7.7	106.8	8.2
Full-scale	97.6	12.9	98.7	11.1	99.5	8.9	98.4	9.3

34.06). All differences between the preoperative and any postoperative scores were highly significant (Table 4). The pre- and postoperative scores showed no correlation.

Both before and after surgery the mother usually rated her child's appearance more critically than the father or the child (Table 5). For both parents, ratings differed significantly between the preoperative assessment and each postoperative evaluation (Table 6). Pre- and postoperative scores correlated only for the mother's pre- versus 2-year postoperative rating ($p < 0.05$).

Discussion

Despite the fact that more than half of our patients had an affected relative, the parents did not forbear to procreate, suggesting a positive attitude and a willingness to live with the handicaps the syndrome entails.

The low preoperative DSM III ratings point to the strains and pressures on the patient and his family at this time, which have a strong effect on their entire well-being. The 1-year postoperative ratings indicate that surgery is seen as a "new lease

on life" and that patients have a feeling of accomplishment and euphoria. The lower 2- and 4-year postoperative scores suggest that they make a more appropriate adjustment when the "honeymoon effect" has worn off and patients realize that they still have some problems.

The patients' higher non-verbal than verbal ability is consistent with findings in other hearing-impaired children (Levine, 1971; Brook and Riggs, 1980). Their self-esteem scores showed the same pattern. As in adaptive functioning, the 4-year postoperative self-esteem scores remained well above preoperative levels.

Table 3 Differences between verbal and performance mean scores and SD for 20 Treacher Collins syndrome patients

Assessment	Difference		Significance (p value)
	Mean	SD	
Preoperative	-9.65	10.14	0.0004
1 year postoperative	-13.15	11.98	0.0001
2 years postoperative	-13.15	9.14	<0.0001
4 years postoperative	-14.75	8.16	<0.0001

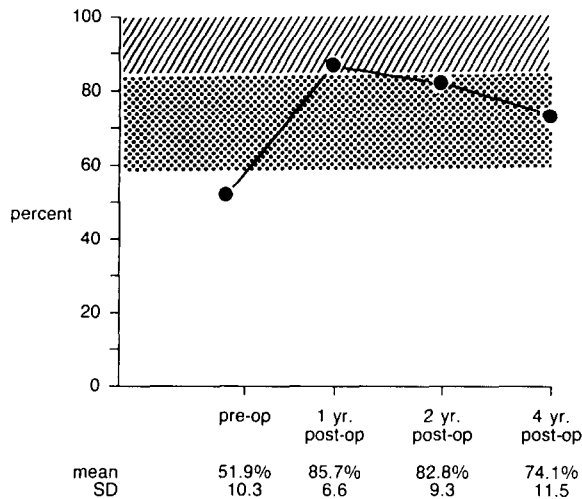


Fig. 2

Figure 2—Pre- and postoperative mean scores and SD on the Piers-Harris Self-Concept Scale for Children. = high, = average, = low

The similarity of scores for patients and parents indicate that the three groups reacted in parallel ways. The fact that the mothers' scores were usually the lowest was also found in an earlier study of the facially disfigured (Lefebvre and Barclay, 1982).

Table 4 Differences between pre- and postoperative ratings of 9 Treacher Collins syndrome patients on the Piers-Harris Self-Concept Scale for Children

Assessments compared with preoperative rating	Difference		Significance (p value)
	Mean	SD	
1 year postoperative	-33.78	10.23	<0.0001
2 years postoperative	-30.89	10.53	<0.0001
4 years postoperative	-22.22	11.34	<0.0004

Conclusions

The findings of this preliminary study suggest that facial surgery is a crucial event in the patients' lives. Their average overall learning potential, which is coupled with elevated non-verbal talents, their familiarity with the syndrome, and the substantial postoperative changes in facial appearance, self-esteem and adaptive functioning affect the quality of their lives positively, giving them hope and encouragement. However, despite its many merits, facial surgery is not a panacea for these patients and their families. A full family assessment is needed before and after surgery. Preoperatively, patients' motivations and expectations for facial reconstruction and the family's planned involvement should be carefully evaluated.

Table 5 Pre- and postoperative ratings of patients' appearance on the Hay's Rating Scale

Rater	Preoperatively			1 yr postoperative			2 yr postoperative			3 yr postoperative		
	Mean	SD	N	Mean	SD	N	Mean	SD	N	Mean	SD	N
Patients	5.1	2.9	17	2.2	1.3	15	3.3	1.1	15	3.7	1.7	15
Mothers	5.9	1.9	19	2.5	0.5	18	3.7	1.4	18	3.6	1.0	18
Fathers	5.3	1.7	14	2.7	0.7	14	3.4	0.9	14	3.3	0.6	14

Table 6 Differences of pre- and postoperative ratings for Treacher Collins syndrome patients on the Hay's Scale of Appearance

Raters	N	Assessment compared with preoperative rating	Difference		Significance (p value)
			Mean	SD	
Patients	15	1 yr postoperative	3.07	2.74	0.0007
		2 yr postoperative	1.93	2.71	0.0153
		4 yr postoperative	1.60	2.97	0.0558
Mothers	18	1 yr postoperative	3.22	1.77	<0.0001
		2 yr postoperative	2.05	1.63	0.0001
		4 yr postoperative	2.11	1.68	0.0001
Fathers	14	1 yr postoperative	2.57	1.55	<0.0001
		2 yr postoperative	1.93	1.73	0.001
		4 yr postoperative	2.00	1.71	0.0007

The limitations of the surgical results should be outlined clearly so that unnecessary disappointment can be avoided. Postoperatively, patients and their families should be helped in adjusting to the new facial image and the life changes beginning at this time.

Long-term follow-up is needed as well. Observations and therapeutic interventions should be continued, even past the initial 4-year postoperative period, and would provide valuable documentation about the psychosocial adjustment during the patient's life cycle. Such data would entitle us to speak with more authority about the patient with Treacher Collins syndrome and develop more expertise in helping him come to terms with his face.

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