



Abstract 72: Long-term Functional Upper Extremity Outcomes in Adult Apert Syndrome Patients

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RESULTS: Sixty-four women were included for analysis (CPM $n=28$ (44%), unilateral $n=36$ (56%)). Mean age of the two groups differed significantly (CPM 44 ± 9 ; unilateral 52 ± 8 ; $p < 0.001$). Mean BMI, race/ethnicity (overall 84% Black or Hispanic), education, income, and mastectomy weight did not differ between groups. Women in CPM group had higher rates of adjuvant therapy (CPM=36% both chemo and radiation, unilateral=6%; $p=0.02$). The rate of having any postoperative complication was higher in CPM group (CPM=50%, unilateral=25%; $p=0.04$). The median postoperative survey time was 11.8 (IQR 6–13) months. For satisfaction with breasts, mean postoperative-preoperative score difference varied significantly between groups, with CPM group having higher and unilateral group having lower satisfaction in the postoperative period relative to preoperative assessment (CPM $= 7.5 \pm 27$, unilateral $= -11.4 \pm 26$; $p < 0.01$). Median postoperative scores for satisfaction with breasts (CPM = 58 [IQR 53–65.5], unilateral = 54.5 [IQR 39.5–61.5]; $p=0.13$), satisfaction with outcome (CPM = 67 [IQR 55–75], unilateral = 74 [IQR 55–86]; $p=0.45$), psychosocial well-being (CPM = 55 [IQR 44.5–73], unilateral = 65 [IQR 41–86]; $p=0.53$), and sexual well-being (CPM = 44 [IQR 26–58], unilateral = 50.5 [IQR 32–56.5]; $p=0.52$) did not differ between groups.

DISCUSSION: Patients electing for CPM are younger and have more advanced disease than those who chose unilateral mastectomy with reconstruction. Despite higher rates of complications in patients undergoing CPM, improved satisfaction with breasts postoperatively may reflect greater satisfaction with breast symmetry.

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Long-term Functional Upper Extremity Outcomes in Adult Apert Syndrome Patients

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PURPOSE: Currently, limited data exist on long-term functional outcomes for patients with Apert syndrome, a rare congenital condition characterized by craniofacial anomalies and complex syndactyly of hands and feet. The study aimed to evaluate upper extremity function and health-related quality of life (HRQOL) in adult Apert syndrome patients.

METHODS: Following Institutional Review Board approval, medical records were queried for patients between ages 18 and 65 years old with Apert syndrome. Recruitment was conducted via postal mail, followed by telephone calls. After obtaining consent, demographic and clinical data, including syndactyly type, were collected during visits and from medical records. Additional patient-reported demographic data were collected through a brief interview. Participants completed two self-reported HRQOL instruments: the 36-Item Short Form Health Survey Instrument, version 2 (SF-36v2), and the Disabilities of the Arm, Shoulder, and Hand (DASH) Outcome Measure. Functional outcomes included moving two-point discrimination, metacarpophalangeal joint range of motion, pinch strength, and the Jebsen Hand Function Test (JHFT).

RESULTS: Between July 2016 and September 2017, 22 participants completed the study. Fifty-five percent of participants were male, 91% were Caucasian, and age ranged from 18 to 43. Sixty-two percent were right-hand dominant and 36% had one digit amputated. Average SF-36 scores were 50.41 ± 12.03 for mental health (best possible score = 100) and 52.77 ± 7.14 for physical health (best possible score = 100). Average total DASH score was 17.73 ± 14.11 (best possible score = 0). Average total JHFT scores for dominant hand were 69.2 seconds for males and 64.7 seconds for females; averages for non-dominant hand were 87.3 for males and 87.7 seconds for females. Average lateral and chuck pinch strengths were 6.98 ± 2.72 and 5.08 ± 2.02 kilograms for dominant hand, respectively, and 6.54 ± 2.53 and 5.33 ± 1.98 kilograms for non-dominant hand, respectively. Average two-point discrimination was 3.95 ± 0.99 millimeters. Average total metacarpophalangeal joint range of motion was 48.60 ± 21.10 degrees. Interviews were completed by all 22 participants. Most subjects reported completing high school, several had completed college and several were considering or enrolled in graduate school. All reported being employed or volunteering at some point during their adulthood. Participants reported living situations that ranged from living with immediate family or a spouse, to living alone.

CONCLUSION: This study offers novel data on long-term functional upper extremity and HRQOL outcomes

in adult Apert patients. Average total scores for the SF-36 mental health, SF-36 physical health, and DASH were higher than population norms. Participants reported a range of paid or unpaid work experience, education, and living situations. Average total JHFT times were slower than population norms, irrespective of hand dominance or sex. Self-reported HRQOL outcomes were more favorable

than functional measures alone would suggest. While these results contribute to meeting a gap in the current literature on Apert patients, a larger cohort would increase understanding of long-term outcomes.

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