

## **Quality of Life in Pediatric Patients After Cone Reconstruction for Ebstein Anomaly**

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### **Introduction:**

Cone reconstruction (CR) has become the standard of care for Ebstein anomaly and has excellent outcomes. However, quality of life (QOL) in patients with Ebstein anomaly after CR has not been reported. Here we sought to evaluate the health status and QOL of pediatric patients who had CR for Ebstein anomaly.

### **Methods:**

We reviewed all patients who had CR from 2007 to 2016 at our institution. Prospective surveys were mailed to all eligible patients. QOL was assessed using the PedsQL 4.0 Generic Core Scales, including four domains: physical, emotional, social, and school functioning.

### **Results:**

Of 116 eligible patients, 71 (61%) responded. Ninety-six percent reported their health as excellent or good, and 55% were symptom free. Sixty-two percent were taking no medications; the most common medication was aspirin in 30%. Only 18% had been hospitalized for cardiac reasons following CR. The average self-reported QOL was 87.6/100 while the average parent proxy-reported QOL was 81.9/100. For the majority, there was no difference by self or parent proxy-report in QOL between CR patients and healthy children; however, QOL was significantly better for CR compared to children with chronic health conditions (Table). By self-report and parent proxy-report, 17% and 15% of patients were deemed “at risk” for reduced QOL respectively. In terms of social functioning, 70/71 (98%) of patients are either full-time students or working full-time.

### **Conclusions:**

Overall, children with Ebstein anomaly who had CR have excellent QOL comparable to healthy peers and significantly better than other children with chronic health conditions. Families of children with Ebstein anomaly can expect excellent QOL, long-term health status and social functioning following CR.

**Table:** PedsQL 4.0 Generic Core Scales for children after cone reconstruction, healthy children, and children with a chronic health condition

	Cone Reconstruction	Healthy Sample	Chronic Health Condition*	<i>p</i> value**	<i>p</i> value***
<b>Patient Self Report</b>					
Total Score	85.57 (14.58)	83.91 (12.47)	74.16 (15.38)	<i>p</i> = 0.22	<b><i>p</i> &lt;0.01</b>
Physical	87.57 (14.94)	87.77 (13.12)	79.47 (17.07)	<i>p</i> = 0.42	<b><i>p</i> &lt;0.01</b>
Psychosocial	84.24 (18.33)	81.83 (13.97)	71.32 (17.13)	<i>p</i> = 0.11	<b><i>p</i> &lt;0.01</b>
Emotional	80.68 (22.29)	79.21 (18.02)	69.32 (21.36)	<i>p</i> = 0.35	<b><i>p</i> &lt;0.01</b>
Social	87.88 (13.78)	84.97 (16.71)	76.36 (21.57)	<b><i>p</i> = 0.01</b>	<b><i>p</i> &lt;0.01</b>
School	82.97 (18.33)	81.31 (16.09)	68.27 (19.05)	<i>p</i> = 0.08	<b><i>p</i> &lt;0.01</b>
<b>Parent Proxy Report</b>					
Total Score	81.85 (15.26)	82.29 (15.55)	73.14 (16.46)	<i>p</i> = 0.75	<b><i>p</i> &lt;0.01</b>
Physical	86.57 (16.16)	84.08 (19.70)	76.99 (20.20)	<b><i>p</i> = 0.03</b>	<b><i>p</i> &lt;0.01</b>
Psychosocial	79.27 (16.46)	81.24 (15.34)	71.04 (17.32)	<i>p</i> = 0.58	<b><i>p</i> &lt;0.01</b>
Emotional	77.55 (21.43)	81.20 (16.40)	71.08 (19.75)	<i>p</i> = 0.24	<b><i>p</i> = 0.02</b>
Social	82.45 (17.86)	83.05 (19.66)	75.06 (21.75)	<i>p</i> = 0.93	<b><i>p</i> = 0.02</b>
School	77.80 (20.76)	78.27 (19.64)	65.58 (20.75)	<i>p</i> = 0.91	<b><i>p</i> &lt;0.01</b>

\* Children with asthma, diabetes, attention deficit hyperactivity disorder, depression, or “other”

\*\* *P* value comparing cone reconstruction cohort to healthy control cohort

\*\*\* *P* value comparing cone reconstruction and children with chronic health conditions