

Effect of bariatric surgery in an Arab child with melanocortin-4 receptor mutation

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Melanocortin 4 (MC4R) receptor mutations are the commonest of the monogenic forms of obesity. We report two cases; siblings with morbid obesity and diabetes mellitus whom underwent bariatric surgery and were later confirmed to have MC4R mutations. Both had achieved developmental milestones normally but progressively gained weight after birth. They were unresponsive to conventional treatment since age three.

A twelve year old boy (weight 143.4 kg, BMI 52 kg/m and HbA1c 8.1 %) diagnosed with diabetes at the age of eleven, had a birth weight of 3.5 kg. The second case, his fourteen year old sister who also had morbid obesity (weight 150 kg, BMI 60-initial and HbA1c not available) developed diabetes at the age of thirteen. Both parents have type 2 diabetes mellitus and are unrelated.

On examination there were no dysmorphic features, obesity was generalized. Of note, were the presence of acanthosis nigricans and mild hypertension. Otherwise general medical examination was unremarkable. Investigations including urea & electrolytes, thyroid and liver function tests were normal. GAD & IA2 antibodies were negative.

Both siblings had sleeve gastrectomy in another centre a week after assessment at ICLDC. The results of genetic testing later revealed a mutation of the MC4R (homozygous) in both of them. Parents showed to be heterozygous for the same mutation.

Within nine months of surgery, the boy had lost 18 kg and reduction in HbA1c to from 8.1% to 6.1 %. The girl had weight reduction of about 22 kg. Unfortunately, data for HbA1c was not available pre surgery for comparison. The girl was losing weight gradually as well as the boy. However, weight loss for the boy was not well maintained. Interestingly, HbA1c levels for the boy were reduced in parallel with weight but for the sister HbA1c levels were increasing after the surgery.

To our knowledge this is the first description of bariatric surgery in children with MC4R mutation in UAE. These cases are unique in demonstrating that sleeve gastrectomy may have different outcomes even in different members of the same family with the same MC4 mutation. This may have major implications for other such patients. It may also provide insights into the hormonal/biological pathways implicated in bariatric surgery.

