Anaesthetic challenges in staged correction of Andy Gump deformity in a young girl with severe obstructive sleep apnoea

Sir,

Patients with Andy Gump deformity have the appearance of absent chin due to an anterior mandibular defect with an extremely retrognathic mandible.[1,2] No report of congenital Andy Gump deformity is available to the best of our knowledge. Andy Gump is a famous cartoon character, in a popular comic strip, The Gumps, published in the American newspapers. He was depicted to be chinless with an oversized moustache, prominent nose, bald head and his mouth appeared as a small hole in his neck.[1]

A 15-year-old girl presented with snoring, difficulty in eating and inability to open mouth since she was an infant. She was found to have bilateral congenital temporomandibular joint (TMJ) ankylosis with micrognathia, hypoplastic and retrognathic mandible [Figure 1]. She was labelled as having congenital Andy Gump deformity and planned for staged reconstruction. Initial stage involved mandibular osteotomy and bilateral placement of mandibular distractors. Interpositional arthroplasty, condylectomy and coronoidectomy, removal of distractors was for the second stage.

At her first surgery, she weighed 27 kilograms with nil mouth opening and difficulty in speaking. Her heart rate was 126/minute with a systolic murmur. She had haemoglobin of 10 gm/dl and sinus tachycardia. Mitral valve prolapse was observed on echocardiography with eccentric mitral regurgitation (MR), mild tricuspid regurgitation (TR) and a pulmonary arterial systolic pressure (PASP) of 34 mm Hg. On polysomnography, apnoea-hypopnoea index was 35.7 [suggestive of severe obstructive sleep apnoea (OSA)], and minimum peripheral oxygen saturation (SpO_2) during sleep was 78% (for 14 seconds).

In view of nil mouth opening, fibreoptic bronchoscope-guided nasal intubation was performed with dexmedetomidine sedation (1 μg/kg loading dose over 10 minutes followed by 0.5 μg/kg/h) and spray-as-you-go with 2% lignocaine (3 ml). Thereafter, standard induction and maintenance of anaesthesia was done. Intraoperative course was uneventful and in view of severe OSA and prolonged surgery, she was electively ventilated overnight in an intensive care unit (ICU) and extubated the next day. The postoperative course was smooth and she was discharged after 7 days.

Second surgery was done six months later. Now she weighed 33 kilograms and had no mouth opening. Her heart rate was 96/minute and PASP was 30 mm Hg. Polysomnography findings had improved drastically with an apnoea-hypopnoea index of 2.1 (No OSA), minimum SpO_2 during sleep was 85%. As before, fibreoptic bronchoscope-guided nasal intubation was performed with dexmedetomidine sedation. Intraoperative course was uneventful, and a mouth opening of one and a half fingers was achieved postoperatively. The patient was extubated in the operating room at the end of the surgical procedure. She was subsequently discharged from the hospital after 10 days. On follow-up, the patient was doing well with improved nourishment, had joined school and had no snoring while asleep.

Airway management challenges due to TMJ ankyloses are further complicated by associated co-morbidities.[3,4] Children who have congenital TMJ ankylosis or develop it before the age of 5 years are more prone to develop facial deformities.[5] Due to

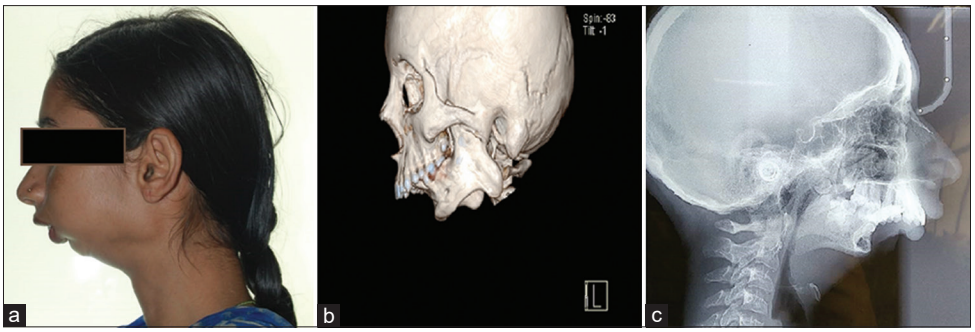

Figure 1: (a) Clinical appearance; (b) three-dimensional radiological image; (c) lateral x-ray

delayed surgery, our patient has severe retrognathia and hypoplastic mandible, characteristic of Andy Gump deformity.

The two-staged correction had different set of challenges before each surgery. Initially, she was malnourished and the airway scenario was difficult with severe OSA. Six months later, though she still had nil mouth opening, she had gained weight with relief in OSA and could be extubated safely after surgical procedure.

A proper assessment of patient with due attention to the inherent challenges is the key to successful management of patients with rare disorders. Andy Gump deformity has the classic triad of Pierre-Robin syndrome (PRS). In contrast to Andy Gump, PRS is most associated with cleft palate (80%) and shows familial association. Also, children with PRS usually get better symptomatically with conservative management (rarely definitive surgery is required) by the age of 2 years, as the structures attain maturity. Although facial morphology might remain altered, respiratory issues and feeding resolve with time.[6]

In conclusion, we wish to highlight that information related to rare disorders like Andy Gump deformity should be shared to assist future caregivers in planning their actions. Also, for the staged surgical procedures, factors pertaining to each stage should be appropriately addressed.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the legal guardian has given his consent for images and other clinical information to be reported in the journal. The guardian understands that names and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

Swati Chhabra, Lovepriya Sharma, Sadik Mohammed, Darshana Rathod

Department of Anaesthesiology and Critical Care, All India Institute of Medical Sciences, Jodhpur, Rajasthan, India

Address for correspondence:
Dr. Swati Chhabra,
Department of Anaesthesiology and Critical Care, All India Institute of Medical Sciences, Jodhpur - 342 005, Rajasthan, India.
E-mail: swati_virgo83@yahoo.co.in

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