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LETTER TO THE EDITOR

Management of a 10-month-old child with a rare combination of Bardet–Biedl syndrome and ano-rectal malformation undergoing anterior sagittal ano-rectoplasty

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To the Editor:

Bardet–Biedl syndrome (BBS) is a rare autosomal-recessive disorder associated with great genetic heterogeneity [1]. Literature describing the anesthetic management of BBS is scanty [2].

A 10-month-old female child born to parents with second degree consanguinity was referred to us with an abnormal site of anal opening and excessive weight gain. She was their first child, whose birth weight was 3 kg. Examination showed a mildly dysmorphic face, short neck, central obesity, polydactyly (six digits) of all four limbs, and partial syndactyly (between the third and fourth finger) of the right hand (Fig. 1). Perineal examination revealed a vestibular fistula, poorly developed labia, and normal vaginal and urethral openings. Her weight was 14 kg (expected weight, $\approx$8.5 kg), length 65 cm, and body mass index 33.1 kg/m^2; milestones were delayed. Cardiovascular and respiratory systems were normal. An ophthalmic examination showed a normal fundus. Laboratory parameters were within normal limits. Echocardiography ruled out any cardiac anomalies. Ultrasonography of the abdomen and pelvis showed a normal renal system, hypoplastic fallopian tubes, and a rudimentary uterus.

A difficult airway cart was kept ready. Anesthesia was induced with sevoflurane in 100% O_2. Intravenous access was secured in the left upper limb. Mask ventilation required a Guedel's airway. Intravenous fentanyl 25 μg was administered, and the trachea was intubated with a 4-mm uncuffed endotracheal tube (laryngoscopic view-2A). Injection of atracurium 7 mg was administered following endotracheal intubation. Caudal analgesia was administered with 10 ml 0.25% bupivacaine. Anesthesia was maintained with O_2 and N_2O (50:50) with sevoflurane (1–2%) with mechanical ventilation (pressure control mode with positive end-expiratory pressure). Vitals were stable, and oxygen saturation was maintained above 95% intraoperatively. She underwent anterior sagittal ano-rectoplasty in lithotomy position uneventfully.

At the end of surgery, 10 ml 0.125% bupivacaine with 15 μg clonidine was administered caudally for postoperative analgesia. Following reversal of neuromuscular blockade and return of consciousness, the trachea was extubated in the operation theatre. Perioperative fluid administration was closely monitored. She had an uneventful postoperative recovery.

BBS is an autosomal-recessive disease characterised by obesity, learning disabilities, dysphormic extremities (syndactyly, brachydactyly, or polydactyly), retinal dystrophy or pigmentary retinopathy, hypogonadism or hypogenitalism (frequently in males), and structural abnormalities or functional impairment of kidneys [3]. If spasticity and mental retardation are present, the criteria are fulfilled for Laurence–Moon–Bardet–Biedl syndrome [4]. Other associated minor clinical signs include diabetes, hypertension, congenital cardiopathy, or Hirschsprung disease [1].

Obesity is a major feature of BBS (72–96% incidence), which usually begins in childhood with the majority of cases exhibiting symptoms within the first year of life. Weight gain, renal function impairment, and retinal degeneration are age-dependent factors [2]. Our patient had obesity, developmental delay, polysyndactyly, and a rare

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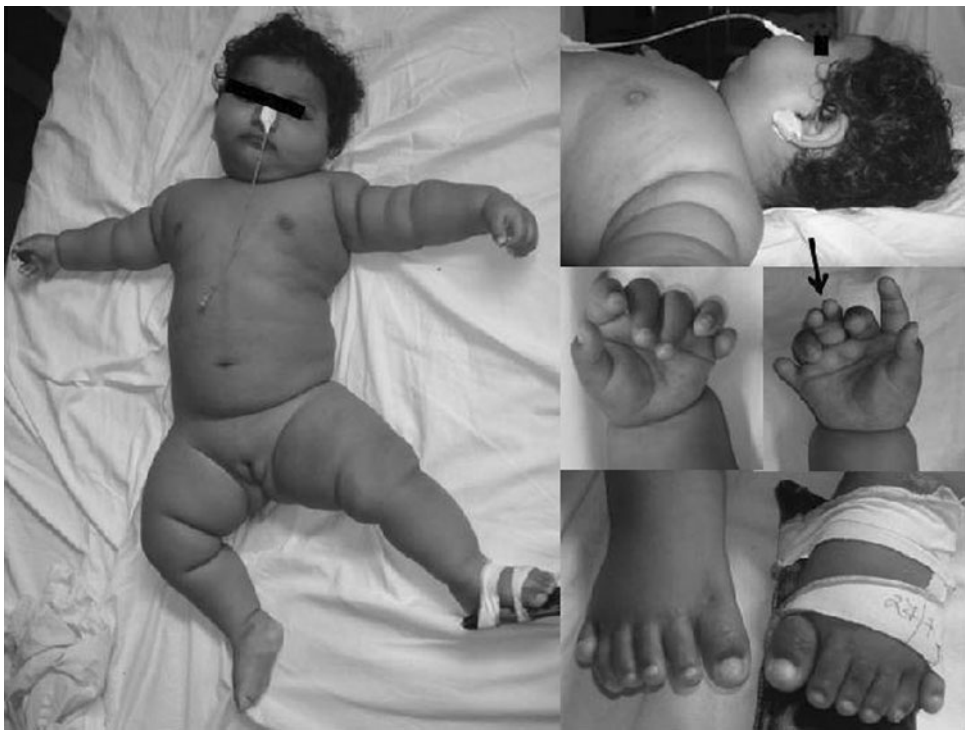

Fig. 1 Appearance of the patient. Mildly dysmorphic face, short neck, central obesity, polydactyly (six digits) of all four limbs, and partial syndactyly (between the third and fourth finger) of the right hand (*arrow*)

combination of BBS and ano-rectal malformation. Various cardiac involvements such as congenital heart defects, hypertrophy of the interventricular septum, and dilated cardiomyopathy have been reported in up to 50% of patients with BBS [5]. Renal failure is the leading cause of death, and survival is substantially reduced.

Anesthetic implications in our patient were related to obesity, difficult venous access, difficult airway, lithotomy position, and avoiding nephrotoxicity. We administered caudal bupivacaine with clonidine for postoperative analgesia. Among the neuraxial adjuvants, clonidine offers clear advantages because of minimal adverse cardiorespiratory effects. To our knowledge, this is the first report describing the anesthetic management of an infant with BBS.

This child is expected to present for multiple surgical procedures and anesthesia later in her life. An uneventful perioperative course of such a patient does not guarantee that the future anesthetics in this patient will be safe and uneventful, as some of the complications such as obesity, airway difficulty, diabetes, renal impairment, and cardiomyopathy associated with BBS are known to worsen with age [2]. Hence, it is emphasized here that when such patients present for anesthesia in the future, they must undergo complete evaluation guided by the clinical status along with a review of the previous anesthetics.

Conflict of interest There are no financial interests and conflicts of interest to declare.

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