Pediatric Anesthesia

Pediatric Anesthesia ISSN 1155-5645

ORIGINAL ARTICLE

Anesthetic considerations for patients with Bardet–Biedl syndrome: a case series and review of the literature

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What is already known

- Bardet–Biedl syndrome is a rare genetic disorder with primary and secondary features affecting multiple organ systems.

What this article adds

- This article highlights the high frequency of airway challenges, cardiac anomalies and dysfunction, and other pertinent disease manifestations to the anesthesiologist.

Keywords
anesthesia; airway management; Bardet–Biedl syndrome; cardiovascular abnormalities; heart defects; congenital; kidney failure; chronic

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Section Editor: Francis Veyckemans

Accepted 16 December 2015

doi:10.1111/pan.12848

Summary

Background: Bardet–Biedl syndrome (BBS) is a rare genetic condition with manifestations that can impact anesthetic and perioperative care. There is a void of literature describing the perioperative anesthetic management in this complex patient population.
Objectives: The purpose of this retrospective series was to describe the perioperative care of patients diagnosed with BBS at a large academic tertiary referral center with experience in caring for these patients.
Methods: All patients receiving anesthesia during the time between July 10, 1997 and Jan 1, 2015 were identified. Anesthetic and perioperative records were reviewed in detail for demographic, echocardiographic, preoperative, intraoperative, and postoperative data.
Results: We identified 12 patients with BBS undergoing 40 anesthetics at our institution. The study identified a high risk for difficult airway with need for advanced airway techniques (67% of patient's ≥18 years of age required either awake fiberoptic or video laryngoscopy techniques), cardiac abnormalities, renal impairment, morbid obesity, and intellectual disability as the main syndrome manifestations of interest to the anesthesiologist. No patient had perioperative complications directly related to BBS; however, each underwent thorough perioperative evaluation with emphasis on the systemic comorbidities associated with the syndrome. This report illustrates that patients with BBS can safely undergo anesthesia, but a detailed and often multidisciplinary preoperative evaluation is prudent.

Introduction

Bardet–Biedl syndrome (BBS), previously known as Laurence–Moon–Bardet–Biedl (LMBB) syndrome and Laurence–Moon syndrome (LMS), is an autosomal recessive and genetically heterogeneous condition with manifestations affecting multiple organ systems (1–5). BBS is now recognized as an isolated syndrome characterized by primary and secondary features (Table 1) (5).

Life expectancy in patients with BBS is significantly reduced and they often present for multiple surgical procedures requiring anesthesia care. Aside from very few

isolated case reports (6–12), there is a void of literature describing the perioperative anesthetic management in this complex patient population. The purpose of this retrospective series was to describe the perioperative care of patients diagnosed with BBS at a large academic tertiary referral center with experience in caring for these patients with special focus on BBS disease manifestations of interest to the anesthesiologist, namely, risk for difficult airway, cardiovascular abnormalities, renal impairment, obesity, and a range of intellectual capacities that can make perioperative care challenging. Although a rare syndrome, review of the pertinent clinical features of BBS for anesthesiologists tasked with caring for these patients is needed. This serves as the largest series detailing perioperative and anesthetic care for patients with BBS.

Materials and methods

After Institutional Review Board approval, all patients with a confirmed diagnosis of BBS within the electronic medical record between July 10, 1997 and Jan 1, 2015 were identified. In accordance with Minnesota state law, only patients authorizing use of their medical records for research purposes were included (historically > 96% of patients at our institution). Only patients requiring anesthesia were included in the study.

Preoperative demographics

The medical and anesthetic records were reviewed for demographic, laboratory, echocardiographic, cardiac catheterization, anesthesia, and operative data. Laboratory data were analyzed for most recent preanesthetic estimated glomerular filtration rate (eGFR) which was calculated from serum creatinine using the isotope dilution mass spectrometry (IDMS) traceable Modification of Diet in Renal Disease (MDRD) study equation. Body mass index (BMI) was calculated and categorized based on the World Health Organization recommendations.

Table 1 Diagnostic criteria of Bardet–Biedl syndrome[a]

Primary features	Secondary features
Obesity	Left ventricular hypertrophy
Renal anomalies	Congenital heart disease
Retinal (rod/cone) dystrophy	Hepatic fibrosis
Postaxial polydactyly	High arched palate/dental crowding/small teeth
Learning disabilities	Diabetes mellitus
Hypogonadism	Nephrogenic diabetes insipidus
	Speech disorder
	Strabismus/cataracts/astigmatism
	Brachydactyly/syndactyly
	Developmental delay
	Ataxia/poor coordination/imbalance
	Mild spasticity (especially lower limbs)

[a]Modified diagnostic criteria: 4 primary or 3 primary + 2 secondary features. Adopted with permission from Beales, P. L., N. Elcioglu, *et al.* (5).

Echocardiographic data

Preoperative transthoracic echocardiograms within 1 year prior to anesthetic delivery were reviewed for left ventricular ejection fraction (LVEF), left ventricular (LV) size, LV mass quantification, right ventricular (RV) size and function, valvular abnormalities ≥ moderate in severity, and congenital cardiac abnormalities. Left ventricular mass quantification was classified as normal, concentric remodeling, and concentric or eccentric hypertrophy based on American Society of Echocardiography's guidelines. The most recent echocardiogram performed on patients who had >1 anesthetic within 1 year were recorded as separate studies and data were analyzed on a per anesthetic basis.

Intraoperative findings

Intraoperative records were reviewed for presence of refractory hypotension (systolic blood pressure [SBP] <90 mmHg or mean arterial pressure <60 mmHg in patients >10 years of age, SBP <60 in patients <1 year of age, SBP 70+2*age in years in patients 1–10 years of age, for >10 min), hypoxemia (SaO2 < 88% for >5 min), ventricular tachycardia/fibrillation or clinically significant dysrhythmia (requiring cardioversion/defibrillation or new antiarrhythmic medications) noted in the medical record, cardiac arrest, or aspiration.

Postoperative outcomes

Postoperative records were reviewed for 30-day complications including aspiration, pneumonia, unanticipated delay of extubation, new renal failure requiring dialysis, acute coronary syndrome, clinically significant dysrhythmia, acute congestive heart failure requiring medical therapy, and cardiac arrest, as documented as diagnoses in the medical record. Thirty-day mortality was recorded.

Literature summary

To enrich and inform the finding here, we conducted a literature review for syndrome manifestations, anesthetic management, and operative care of patients with

BBS. A search was performed in Ovid MEDLINE (1946–Jan. 2015), Ovid Embase (1988–Jan. 2015), and the Web of Science (1975–Jan 2015). The search strategy included MeSH terms (Bardet–Biedl Syndrome) and keywords, such as Laurence–Moon–Bardet–Biedl syndrome, Laurence–Moon–Biedel syndrome and anesthesia, all limited to the English language. Bibliographies of published literature were analyzed for additional references and cases describing perioperative care were reviewed for complications.

Results

A review of the electronic medical records database identified 12 patients with BBS who underwent a total of 40 anesthetics at our institution. Each patient was evaluated on a per anesthetic basis as the disease manifestations of BBS progress over time making subsequent anesthetics in the same patient variable due to changing patient comorbidities. The demographic and perioperative data are summarized in Table 2.

Preoperative demographics

Twenty anesthetics (50%) occurred in patients <18 years of age. Renal impairment secondary to BBS was present prior to eight anesthetics (one renal transplantation operation, four with a history of renal transplantation, and three dialysis-dependent renal failure). Twelve anesthetics were conducted on patients with obstructive sleep apnea, and four occurred in patients with prior gastric bypass operations for obesity.

Echocardiographic data

Preoperative transthoracic echocardiograms within 1 year of the operative date were completed prior to 29 of 40 anesthetics, with the majority, 19 (66%), showing cardiac abnormalities (Table 3). Of the seven anesthetics with left ventricular concentric hypertrophy, five (71%) were <18 years of age and all seven had associated congenital heart disease. Of the five anesthetics in patients with left ventricular eccentric hypertrophy, all five were ≥18 years of age. The overall mean left ventricular ejection fraction (LVEF) was 57.2%. The mean LVEF among patients >18 years of age was 41.7% (median 51%, SD ±19.7%, range 10–60%). Of 23 echocardiograms reporting right ventricular systolic pressure (RVSP), eight (35%) had RVSP values >35 mmHg. Of those with RVSP >35 mmHg, the mean systolic blood pressure was 117 mmHg. Two anesthetics had preoperative right heart catheterization with a mean PAP of 40 ± 11 mmHg (median 39.5 mmHg, range of 32–47 mmHg). Among the cases of pulmonary hypertension, six (67%) were <18 years of age (mean 17.9 years of age, SD ± 13.0, range of 4–42 years of age).

Table 2 Demographic and perioperative data

Anesthetics	40
Gender	M = 24 (60%)
Age (years)	Mean = 21.6
	Range = 6 month–56
	<10 years: 7 (18%)
	11–17 years: 13 (32%)
	≥18 years: 20 (50%)
	Median = 17
	SD ± 15.3
BMI ($kg \cdot m^{-2}$)	Mean = 37.9
	<10 years: mean BMI 28.2
	11–17 years: mean BMI 33.6
	≥18 years: mean BMI 43.0
	Range = 18.9–55.7
	Median=38
	SD ± 9.5
eGFR[a] ($ml \cdot min^{-1}$)	Mean = 48.6
	Range = 5–>60
	Median = 60
	SD ± 19
ASA classification[b]	ASA I = 2 (5%)
	ASA II = 13 (35%)
	ASA III = 19 (51%)
	ASA IV = 3 (8%)
Procedure	Urogenital = 12 (30%)
	Orthopedic = 9 (22.5%)
	Cardiac = 8 (20%)
	ENT = 6 (15%)
	Gastrointestinal = 3 (7.5%)
	Vascular = 2 (5%)
Anesthesia type	General = 36 (90%)
	MAC = 2
	Regional = 1
	Nurse directed sedation = 1
Airway	Direct laryngoscopy = 21 (52.5%)
	Awake fiberoptic = 6 (15%)
	Videolaryngoscope = 4 (10%)
	Laryngeal mask airway = 4 (10%)
	None (e.g., MAC, regional, moderate sedation) = 4 (10%)
	Mask with Parson's laryngoscope = 1[c](2.5%)

ASA, American Society of Anesthesiologists; BMI, body mass index; DL, direct laryngoscopy; eGFR, estimated glomerular filtration rate; ENT, ear, nose and throat; ETT, endotracheal tube; F, female; M, male; MAC, monitored anesthesia care; *n*, number; OTT, oral tracheal tube; SD, standard deviation.

[a]Normal eGFR at the Mayo Clinic Laboratory was reported as >60 $ml \cdot min^{-1}$.

[b]ASA classification was only documented in 37 anesthetic records.

[c]Performed during sleep state laryngoscopy, microlaryngoscopy, and bronchoscopy.

Intraoperative findings

A variety of operations were performed with the most common being urogenital (30%), orthopedic (22.5%), cardiac (20%), and ear, nose, and throat (ENT) (15%) (Table 2). Of these procedures, 28 (70%) were performed for disease manifestations of BBS. No operations were performed emergently. Of 32 tracheal

Table 3 Preoperative echocardiography data

Preoperative transthoracic echocardiograms[a]	29
Left ventricular ejection fraction[b]	Severe hypokinesis (EF<20%) = 2 (7%)
	Moderate hypokinesis (EF 21–40%) = 2 (7%)
	Mild hypokinesis (EF 41–55%) = 4 (15%)
	Normal (56–70%) = 15 (56%)
	Hyperdynamic (EF>71%) = 4 (15%)
	Mean = 57.2%
	Median = 63%
	SD ± 18.4
	Range = 10–78%
Left ventricular size	Normal = 22
	Severe enlargement = 4
	Mild enlargement = 3
Left ventricular mass quantification[c]	Concentric hypertrophy = 7
	Eccentric hypertrophy = 5
	Concentric remodeling = 9d
	Normal = 4
	Not reported = 4
Right ventricular systolic pressure[e]	Normal = 15
	Elevated (RVSP >35 mmHg) = 8
	Mean 36.9 mmHg
	Median = 32 mmHg
	SD ± 15.29 mmHg
	Range = 21–95 mmHg
Right ventricular size[f]	Normal = 25
Right ventricular function[g]	Normal = 28
Valve disease[h]	Moderate MR = 4
	Severe AR, Moderate TR = 1
	Severe AR, moderate AS and moderate MR = 1
Congenital heart disease	AV canal defects *n* = 6
	Repaired AV canal defect = 3
	Repaired AV canal defect with mild LV outflow obstruction = 1
	Repaired AV canal defect with mild LV outflow obstruction and bicuspid aortic valve = 1
	Repaired AV canal defect with severe LV outflow obstruction = 1
	Bicuspid aortic valve = 4
	Repaired = 3
	Native = 1
	Isolated LV outflow tract obstruction = 3
	Mild = 2
	Severe = 1
	Bilateral pulmonary artery stenosis = 2
	ASD = 1

AR, aortic regurgitation; AS, aortic stenosis; ASD, atrial septal defect; AV, atrioventricular; ECHO, echocardiography; LV, left ventricle; LVEF, left ventricular ejection fraction; M, male; MR, mitral regurgitation; *n*, number; SD, standard deviation; TR, tricuspid regurgitation.

[a]Twenty-nine preoperative echocardiograms performed in nine patients. Of 29 examinations 19 in males, mean age 17.5 years of age, median 17 years of age, SD ± 11.1, Range 2 months–42 years of age.

[b]Ejection fraction only reported in 27 preoperative echocardiograms.

[c]Calculation of LV mass based on the American Society of Echocardiography recommendations (28).

[d]Concentric remodeling based on the American Society of Echocardiography recommendations (28) was not documented as an abnormality.

[e]Right ventricular systolic pressure only reported in 23 preoperative echocardiograms.

[f]Right ventricular size only reported in 25 preoperative echocardiograms.

[g]Right ventricular function only reported in 28 preoperative echocardiograms.

[h]Only valvular disease classified as ≥ moderate was included.

intubations, 17 were completed on patients <18 years of age and 15 on patients ≥18 years of age. All 17 tracheal intubations in children were uneventfully completed with direct laryngoscopy. Of the 15 tracheal intubations performed on patients ≥18 years of age, the most common approach was through indirect laryngoscopic techniques with six (40%) completed via awake fiberoptic intubation and four (27%) via video laryngoscopy.

Postoperative outcomes

There were no intraoperative complications documented during the 40 anesthetics. Postoperative complications were documented after three anesthetics in three separate patients. One patient with a history of prior unprovoked DVT and PE had a cardiac arrest on postoperative day (POD) 2 following an orthopedic procedure, and was found to have a massive pulmonary embolism (PE) despite a therapeutic INR of 2.3. The patient expired on POD 6 after care was transitioned to comfort measures. Another patient, on POD 3 following cardiac catheterization and VT ablation, required a short hospitalization at an outside hospital for acute congestive heart failure. A third patient developed a renal hematoma 6 days following renal biopsy requiring a 1-day hospitalization with no subsequent complication.

Literature summary

A comprehensive literature search identified only seven isolated case reports in the English language describing anesthetic and/or airway management in patients diagnosed with BBS (Table 4) (6–12). In these individual case reports, difficult airways, airway abnormalities, and cardiac abnormalities were not infrequent.

Discussion

BBS manifestations often require effected patients to undergo multiple operations throughout their lifetimes. Disease manifestations of importance to anesthesiologists include: risk for difficult airway, cardiac abnormalities, renal impairment, obesity, and a range of intellectual capacities that can make perioperative care challenging (6–12). There is very limited literature describing the perioperative care of these complex patients. The main finding of this study was that BBS patients can safely undergo anesthesia and surgery but frequently require advanced airway management and often have cardiac abnormalities. At our large tertiary care center, patients with BBS were often identified and diagnosed early in life which allowed for detailed and multidisciplinary evaluation of the various manifestations of BBS. These unique manifestations require recognition and appropriate management, as prior uncomplicated anesthetics do not ensure subsequent uneventful perioperative care as BBS manifestations progress over time. Although rare, review of pertinent clinical features of BBS for anesthesiologists tasked with caring for these patients is necessary. This serves as the largest series detailing perioperative and anesthetic care for patients with BBS.

BBS is a well-established clinical syndrome similar to LMBBS and LMS first described by Laurence, Moon, Bardet, and Biedl in the late 19th and early 20th century (1–3). BBS has been differentiated from LMS by the primary and secondary features outlined in Table 1 in addition to the lack of polydactyly seen in LMS (5).

BBS is known to be an autosomal recessive, genetically heterogeneous disorder with 18 described gene mutations accounting for the majority of cases (13). Ciliary defects have been proven to be the mechanism leading to the phenotypes associated with BBS (14). The prevalence of this syndrome varies from one in 13 500 patients of Middle Eastern and Canadian descent to one in 160 000 patients of European descent (15). Other syndromes are indeed very similar to BBS including Alström, Rubinstein–Taybi, Biemond Syndrome Type II, Cohen, and McKusick–Kauffman syndrome in which difficult airway management is also an important consideration (16).

In our study, advanced airway techniques were often necessary as difficult airways were frequently encountered. All tracheal intubations were successfully completed in pediatric patients (<18 years) with direct laryngoscopy. Contrarily, 67% of adults required indirect laryngoscopy techniques (40% awake fiberoptic technique, 27% video laryngoscopy) due to known or anticipated airway difficulties, suggesting airway difficulty becomes more profound with age likely due to worsening oral and facial anomalies in addition to progressive morbid obesity. Of note, video laryngoscopes were introduced to our institution in 2009. Of the four video laryngoscope intubations, all were completed on patients with a prior history of awake fiberoptic intubation. Of awake fiberoptic intubations, two were performed despite video laryngoscope availability. There were no notations of perioperative respiratory complications in this cohort.

Manifestations of BBS that may contribute to airway difficulty include dental anomalies, high arched palate, facial dysmorphism, and morbid obesity (5,6). Bifid epiglottis had not previously been an associated

Table 4 Published case reports of patients with BBS undergoing anesthesia

Author	Age/gender	Surgical procedure	Indication	Cardiovascular abnormalities	Anesthesia type	Airway findings	Airway management
Low and Brown 1992 (12)	10 month M	Muscle biopsy	Family history of death under anesthesia	Not reported	Spinal anesthesia with sedation	Not reported	None
Urben and Baugh 1999 (6)	10 M	ENT airway evaluation	Multiple intubations for respiratory arrest secondary to upper respiratory infection, aspiration pneumonia, prolonged intubation	VSD	General	Bifid epiglottis	DL
Mahajan *et al.* 2007 (7)	32 F	ORIF of the right femur and tibia	Femur and tibial fracture	Dilated cardiomyopathy; EF 30%	Combined spinal/ epidural anesthesia	Anticipated difficult airway with Mallampati III	DL
Chittoodan and Crowe 2010 (8)	8 F	MRI brain and spinal cord	Not reported	Not reported	General	Bifid epiglottis	DL
Katsika *et al.* 2011 (9)	13 F	Renal transplantation	Renal failure	Not reported	General	Bifid epiglottis	DL, Cormack–Lehane grade III airway
Hegde *et al.* 2012 (10)	10 month F	Anterior sagittal ano-rectoplasty	Abnormal anal opening, excessive weight gain	Not reported	Combined general and caudal anesthesia	Dysmorphic facies	DL
Bhat *et al.* 2014 (11)	8 M	Postauricular dermoid excision	Dermoid cyst	Not reported	General	Anticipated difficult airway with Mallampati III	DL

DL, direct laryngoscopy; EF, ejection fraction; ENT, ear nose and throat; F, female; LV left ventricle; M, male; MR, mitral regurgitation; MRI, magnetic resonance imaging; ORIF, open reduction and internal fixation; TEE, transesophageal echocardiogram; VSD, ventricular septal defect.

manifestation of BBS but three individualized case reports describe this incidental finding, with one case describing multiple airway complications (Table 4) (6,8,9). A difficult airway was anticipated preoperatively in three published case reports as evidenced by physical exam or previously known difficult airway (6,7,11). All six published case reports describing airway management for patients with BBS recounted successful direct laryngoscopy; however, these authors reported the immediate availability of additional airway devices when difficulty was anticipated (7,10,11).

Cardiac manifestations of BBS have previously been reported as congenital heart disease, ventricular hypertrophy, and cardiomyopathy (5,17–19). In 1964, McLaughlin, *et al.* evaluated three brothers and reviewed 330 published cases of patients with LMBBS. There were nine cases of congenital heart disease, predominantly in males, including atrial septal defect (ASD), ventricular septal defect (VSD), transposition of the great arteries, pulmonary stenosis, hypoplastic aorta, patent ductus arteriosus (PDA), and dextrocardia. There were seven reports of acquired heart disease including biventricular hypertrophy and left ventricular hypertrophy which were associated with renal disease and systemic hypertension (17). In 1994, Elbedour, *et al.* reviewed 33 patients with BBS from three extended families who underwent 22 echocardiograms. Seven (32%) had echocardiographic abnormalities (three congenital heart disease, two intraventricular hypertrophy, one dilated cardiomyopathy, and one mild-moderate tricuspid regurgitation). The observed cases of congenital heart disease consisted of bicuspid aortic valve, ASD, and pulmonic stenosis. There was no correlation found between cardiac disease, renal dysfunction, and systemic hypertension (18). There have been further case reports of isolated dilated cardiomyopathy in patients with BBS (7,19).

In our study, preoperative echocardiograms within 1 year of the surgical procedure were completed prior to 29 anesthetics, with 19 (66%) of these studies demonstrating abnormalities (Table 3). The overall mean left ventricular ejection fraction LVEF was 57.2%, however, the mean LVEF among patients >18 years of age was 41.7%, emphasizing the progressive nature of cardiac dysfunction over time. Diagnostic signs of pulmonary hypertension were discovered in several patients. Of echocardiograms reporting RVSP, eight (35%) had values >35 mmHg, with a mean systolic blood pressure of 117 mmHg. Right heart catheterization was performed prior to two anesthetics with a mean PAP of 40 ± 11 mmHg. These findings may be explained by the high incidence of OSA in the patient population, along with long-standing cardiac pathology. Overall, the echocardiographic findings suggest that patients with BBS undergoing surgery have a high incidence of congenital heart disease and also acquired heart disease, the latter of which may have anesthetic implications with progression over time.

Renal impairment leading to renal failure can be present in 90% of BBS patients and is the leading cause of death (20,21). Both structural and functional renal anomalies have been described with the syndrome (5). BBS patients requiring renal transplantation at ages ranging from 6 to 58 years have been described in case reports (22,23). Renal transplantation was unremarkable in these patients but many suffered from long-term complications related to glucocorticoid immunosuppression contributing to morbid obesity postoperatively (21,23).

In our study, a wide range of renal function was present in patients undergoing operative care. The preanesthetic mean calculated eGFR was 48.6 $ml \cdot min^{-1}$ (normal >60 $ml \cdot min^{-1}$), correlating to stage 3A chronic kidney disease. There was one anesthetic for renal transplantation at our institution, four anesthetics for patients who had previously undergone renal transplantation, and three anesthetics for patients with dialysis-dependent renal failure. The mean age of renal transplantation of subjects in our study was 18.3 years of age with a range of 7–25 years of age. All the patients in our study underwent successful renal transplantation without complication.

Obesity is another primary feature of BBS with a frequency between 72% and 96%, with rapid weight gain noted in the first year of life (24–26). Interestingly, compared to non-BBS obese patients with similar BMI, adult BBS patients had considerably more visceral fat while pediatric BBS patients appear to have more diffuse adiposity (26,27).

The mean BMI of patients undergoing anesthesia was 38 $kg \cdot m^{-2}$. There were four anesthetics completed on patients who had previously undergone successful gastric bypass related to medically challenging obesity. The higher adult compared to pediatric BMI suggests that these patients become increasingly obese with age, potentially contributing to perioperative risk.

Disease manifestations which may also impact perioperative care include intellectual disabilities, aggressive behavior, liver dysfunction, retinal dystrophy leading to blindness, and diabetes mellitus (6–11). Children with BBS have been described as having labile behavior with elements of obsessive compulsive, autistic spectrum disorder, or even psychosis. Most patients develop visual symptoms prior to teenage years while the majority of patients are legally blind by the second or third decade of life (26). For patients with intellectual disabilities and

aggressive behavior, premedication may be needed to calm patients of varying age as soothing techniques, good patient–physician rapport, and parental assistance may not be sufficient.

Although most patients in this review underwent general anesthesia, there are no inherent contraindications for neuraxial and/or regional techniques related to BBS. Two patients in the literature review underwent neuraxial anesthesia without complication (7,10). Although three patients in this study had postoperative complications (PE POD 2 with subsequent death POD 6, acute congestive heart failure POD 3, and renal hematoma POD 6), none of these appear directly related to BBS.

Limitations of this study include all those inherit to a retrospective series, including charting omissions and the potential for data misinterpretation. Given that many patients present to our tertiary referral center specifically for perioperative care and then receive follow-up care elsewhere, comprehensive 30-day follow-up may be incomplete.

In conclusion, BBS is a rare genetic condition with important implications which may directly impact safe anesthetic and perioperative care. Of importance to anesthesiologists include risk for difficult airways, cardiac abnormalities, renal impairment, morbid obesity, and intellectual disability. Progression of many of these syndrome manifestations over time should caution anesthesiologists to conduct a detailed and often multidisciplinary evaluation regardless of prior uneventful perioperative care. This study revealed that patients with BBS can safely undergo anesthesia, but a thorough preoperative evaluation with attention to airway examination, cardiovascular abnormalities, and preservation of renal function is prudent as the frequency of these abnormalities is significantly more common in this patient population. Although no patients in our review experienced perioperative complications directly related to BBS, the potential for such events should be recognized, and appropriate preventative strategies utilized.

Ethics approval

Institutional Review Board approval was obtained for this study.

Funding

This research was carried out without funding. No authors have financial ties, incentives, or disclosures.

Conflict of interest

No conflicts of interest declared.

Acknowledgments

The authors acknowledge the contribution made by Ann M. Farrell in performing a comprehensive literature search. We acknowledge contributions made in designing and performing the patient data search for this project from Alberto Marqeuz, R.R.T and the Anesthesiology Clinical Research Group at Mayo Clinic, Rochester, MN.

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