A retrospective study to evaluate the anaesthetic choices and complications for patients with osteogenesis imperfecta at a quaternary referral hospital

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Background: Osteogenesis imperfecta (OI) is an inherited genetic syndrome affecting connective tissue. Patients often undergo surgery due to an increased susceptibility to bone fractures. Anaesthesia is associated with many perioperative challenges. This study aimed to describe and evaluate the perioperative management of OI paediatric patients presenting for surgery at Inkosi Albert Luthuli Central Hospital (IALCH).

Methods: A retrospective chart review of children under 18 years who had OI and underwent surgical procedures from 2000 to 2017 at a quaternary referral hospital was conducted. Patients were identified from the electronic patient database. The following variables were extracted: demographic data, preoperative history, examination, investigations, chronic medications, intra- and postoperative management and perioperative complications. Simple descriptive statistics were performed using a Microsoft Excel spreadsheet.

Results: Thirty-nine patients who underwent 93 surgeries were included. The majority (72.1%) had severe type III OI and had elective orthopaedic surgery. Anaemia was identified in 64.5% of patients; 40.8% had a spinal deformity and 37.6% had an abnormality on respiratory examination. A supraglottic airway device (SGAD) was used in 91.9% of patients, with only three airway complications. Eighty-seven per cent of cases had combined general (GA) and regional anaesthesia (RA). No children had documented signs suggestive of hypermetabolism or malignant hyperthermia.

Conclusion: Despite most patients in our study having severe OI, few of the complications and difficulties described in the literature were identified. A combined GA and RA technique with a SGAD was shown to be a safe anaesthesia technique. Improved preoperative investigation, especially a full blood count due to the high incidence of anaemia, should be encouraged to improve overall care.

Keywords: anaesthesia, osteogenesis imperfecta, paediatric perioperative outcomes

Introduction

Osteogenesis imperfecta (OI) is the most common, albeit rare, genetic syndrome affecting connective tissues, occurring in one in 15 000 to 20 000 live births. It is caused by a genetic mutation resulting in abnormal collagen formation or a decrease in the amount of collagen production. The clinical presentation is heterogeneous, depending on the severity. Clinical features include frequent fractures, often after minor trauma, increased joint mobility, reduced bone mass, short stature, progressive skeletal deformities, blue sclerae, abnormal teeth, fragile skin, and adult-onset deafness. The classification of OI has become complex with additional types added as genetic information becomes available. However the simpler classification by Sillence remains clinically useful and was therefore used in our study (Table I).

The management of OI is multidisciplinary and multimodal. Non-surgical treatment involves physiotherapy, bracing, splinting and rehabilitation to strengthen muscle and decrease the risk of positional deformities and fractures. Medical management includes bisphosphonate therapy (BP) and growth hormone (GH) to increase bone strength and decrease the fracture risk.

Surgical treatment is often needed to manage fractures and correct deformities during childhood. These patients pose numerous anaesthesia challenges due to kyphoscoliosis, cardiac and pulmonary disease which could influence anaesthesia management. In addition, they may pose difficulties with airway management. Careful attention needs to be paid to positioning

Table 1: Clinical classification – modified from Sillence et al

| OI type | Severity | Inheritance | Clinical form | Biochemical abnormalities | Gene mutation |
|---------|----------|-------------|---------------|----------------------------|---------------|
| I       | Mild and non-deforming | AD | Mild deformity, variable DI, blue sclera, early deafness | 50% reduction in type I collagen | COL1A1 |
| II      | Perinatal lethal | AD AR (rare) | Extreme bone fragility, short and wide long bones, perinatal death | Structural alteration of type I collagen (glycine substitution) | COL1A1/2 |
| III     | Severe/ progressively deforming | AD AR | Severe bone fragility and early KS | Structural alteration of type I collagen (glycine substitution) | COL1A1/2 |
| IV      | Moderate | AD | Moderate deformity of long bones and spine, short stature, DI | Structural alteration of type I collagen (glycine substitution) | COL1A1/2 |

AD – autosomal dominant, DI – dentinogenesis imperfecta, KS – kyphoscoliosis, AR – autosomal recessive
in the perioperative period to avoid fractures. Patients with OI may also be at an increased risk of bleeding and intraoperative hyperthermia, however the association of OI and malignant hyperthermia has not been clearly established.

The main aim of this study was to describe and evaluate the perioperative management of paediatric patients with OI presenting for surgery at Inkosi Albert Luthuli Central Hospital (IALCH).

Methods

Following ethics approval (Biomedical Research Ethics Committee, University of KwaZulu-Natal BEO45\18), we performed a retrospective clinical audit of children below the age of 18 with OI who underwent surgery at IALCH, a quaternary referral hospital. Patients were identified from the IALCH electronic patient database, using the search term "osteogenesis imperfecta" and then filtered with the terms "surgery" and "age under 18". The period included was 1 January 2000 to 31 December 2017. Once cases were identified from the database, the study variables were extracted onto a data collection tool and then entered into a Microsoft Excel spreadsheet (version 2016) by the principal investigator (PI) and verified by a research assistant.

The following variables were collected: baseline demographic data, preoperative history including OI severity, examination, investigations, and medication including bisphosphonate treatment, intraoperative management and complications, and postoperative placement and complications.

Statistical methods

Categorical data were analysed in Microsoft Excel (v2016), using simple descriptive statistics and presented as number (n) and percentage (%).

Results

The database identified 1 240 visits in patients with OI presenting to IALCH over the 17-year study period (Figure 1). Patients over the age of 18 and patients not presenting for surgery were excluded. This identified 93 surgical visits with a final recruitment of 39 patients.

Demographic data are presented in Table II. The sample consisted of two-thirds (66.7%) female patients. Most of our patients were aged 3–6 years (38.7%) or 7–12 years (43.0%) at the time of their operation. The sample was predominantly Black African (93.5%) and the majority had OI type III (72.1%).

| Sex | By case (n = 93) | By patient (n = 39) |
|-----|----------------|-------------------|
|     | n (%)          | n (%)             |
| Female | –             | 26 (66.7%)        |
| Age   |                |                   |
| 0–2 years | 8 (8.6%)       | –                 |
| 3–6 years | 36 (38.7%)     | –                 |
| 7–12 years | 40 (43.0%)     | –                 |
| 13–18 years | 9 (9.7%)       | –                 |
| Race  |                |                   |
| Black African | 87 (93.5%)  | 37 (94.9%)        |
| Other | 6 (6.5%)       | 2 (5.1%)          |
| OI type |              |                   |
| I     | 0 (0%)         | 0 (0%)            |
| II    | 0 (0%)         | 0 (0%)            |
| III   | 67 (72.1%)     | 25 (64.1%)        |
| IV    | 4 (4.3%)       | 1 (2.6%)          |
| Not recorded | 22 (23.6%)  | 13 (33.3%)        |
| Weight (kg) |            |                   |
| <10   | 20 (21.5%)     | –                 |
| 11–15 | 35 (37.6%)     | –                 |
| 16–20 | 9 (9.6%)       | –                 |
| 21–30 | 14 (15.0%)     | –                 |
| 31–40 | 6 (6.4%)       | –                 |
| >40   | 1 (1.0%)       | –                 |
| Not recorded | 8 (8.6%)   | –                 |
| Type of surgery |            |                   |
| Elective | 91 (97.8%)  | –                 |
| Emergency | 2 (2.2%)    | –                 |
| Severity of OI |            |                   |
| Not recorded | 22 (23.7%)  | 13 (33.3%)        |
| Severe | 71 (76.3%)     | 26 (66.7%)        |
| Family history of OI |            |                   |
| No    | 25 (26.9%)     | 14 (35.9%)        |
| Yes   | 9 (9.8%)       | 5 (12.8%)         |
| Not recorded | 59 (63.4%)  | 20 (51.3%)        |
| Previous surgery |            |                   |
| 0     | 22 (23.7%)     | –                 |
| 1     | 17 (18.3%)     | –                 |
| 2–4   | 26 (27.9%)     | –                 |
| >4    | 28 (30.3%)     | –                 |
| Type of surgery |            |                   |
| Orthopaedic | 89 (95.7%)  | –                 |
| Lower limb |            |                   |
| Bilateral femur | 6 (6.5%)      | –                 |
| Bilateral tibia | 3 (3.2%)      | –                 |
| Unilateral femur | 38 (40.8%)    | –                 |
| Unilateral tibia | 24 (25.8%)    | –                 |
| Upper limb | 3 (3.2%)      | –                 |
| Other orthopaedic | 15 (16.1%)  | –                 |
| Non-orthopaedic | 4 (4.3%)   | –                 |

Figure 1: Patient recruitment analysis
A retrospective study to evaluate the anaesthetic choices and complications for patients with osteogenesis imperfecta at a quaternary referral hospital

The type of OI was recorded in 26 (68.4%) patients; of these all had severe OI and all but one had OI type III.

Ninety-one per cent of patients had their weight recorded at the time of their theatre visit. The children had a wide range of weights: the biggest grouping weighed 11–15 kg (37.6%); twenty-one per cent of the patients weighed less than 10 kg, and only one patient weighed more than 40 kg.

The vast majority of our cases underwent elective surgery (97.8%) and most patients (76.3%) had had previous surgery. The majority of procedures were orthopaedic (95.7%) and were mainly lower limb surgery.

The preoperative investigations are reported in Table III. (Each surgery a patient had is described and recorded; i.e. a patient may be counted more than once if investigations were repeated on separate visits). Ninety-eight per cent of patients had a haemoglobin (Hb) recorded. Less than half these patients had a full blood count done (41.9%), the remainder had a finger-prick Hb only. It was found that 64.5% of patients were anaemic at the time of surgery (IALCH laboratory reference range used). About half the patients had a quantitative platelet count, of which 24.7% were elevated, and none were low. Four patients had platelet function testing, which were all normal. Four patients had international normalised ratios (INR) done, all of which were normal. Approximately 30% (29) of patients had a chest radiograph (CXR), and of those, 58.6% (17/29) had an abnormal finding. Only eight patients had pulmonary function testing done; 75% of these tests were found to be abnormal. A further 11 patients had abnormal cervical-spine X-rays. Few patients had cardiac investigations including electrocardiogram (ECG) or echocardiogram (ECHO) performed.

Preoperative assessments are shown in Table IV. Most patients were on bisphosphonate treatment (78.5%). The airway assessment was unremarkable in most cases; however, many children had spinal deformities (40.8%) and some abnormality on respiratory examination (37.6%). Chronic comorbidities were noted in 23 (24.7%) cases including cerebral palsy, epilepsy and human immunodeficiency virus (HIV), but only 2 (2%) cases presented with acute comorbidities like upper respiratory tract infections. Sedative premedication was prescribed for 76.3% of the children.

The intraoperative anaesthesia conduct is shown in Table V. Most children had an inhalational induction of anaesthesia (84.9%) and had their airway maintained with a SGAD (91.9%). One child was given a muscle relaxant. The majority of cases were done using a combination of general and regional anaesthesia (87.0%). A variety of analgesia was used including opioids, NSAIDs, ketamine and regional analgesia.

### Table III: Preoperative investigations

| Investigation | Normal | Abnormal | Not recorded |
|---------------|--------|----------|--------------|
| **Haemoglobin (Hb)** (n = 93) | | | |
| n (%) | n (%) | n (%) |
| Formal | 33 (35.5%) | 58 (63.7%) | 2 (2.2%) |
| Ward | 33 (35.5%) | 42 (45.2%) | 2 (2.2%) |
| **Platelets** | | | |
| n (%) | n (%) | n (%) |
| Normal | 23 (24.7%) | 23 (24.7%) | 47 (50.5%) |
| Elevated | 39 (41.9%) | 1 (1.07%) | 53 (56.9%) |
| **Platelet function** | | | |
| n (%) | n (%) | n (%) |
| Normal | 4 (4.3%) | 0 (0%) | 89 (95.6%) |
| **LFT** | | | |
| n (%) | n (%) | n (%) |
| Normal | 24 (25.8%) | 0 (0%) | 69 (74.1%) |
| **ECG** | | | |
| n (%) | n (%) | n (%) |
| Normal | 3 (3.2%) | 0 (0%) | 89 (95.6%) |
| **PFT** | | | |
| n (%) | n (%) | n (%) |
| Normal | 2 (2.1%) | 6 (6.4%) | 85 (91.3%) |
| **ECHO** | | | |
| n (%) | n (%) | n (%) |
| Normal | 3 (3.2%) | 0 (0%) | 89 (95.6%) |
| **CXR** | | | |
| n (%) | n (%) | n (%) |
| Normal | 12 (12.9%) | 17 (18.2%) | 64 (68.8%) |
| **TFT** | | | |
| n (%) | n (%) | n (%) |
| Normal | 5 (5.3%) | 0 (0%) | 88 (94.6%) |
| **ABG** | | | |
| n (%) | n (%) | n (%) |
| Normal | 9 (6.9%) | 2 (2.1%) | 82 (88.1%) |
| **C-spine X-ray** | | | |
| n (%) | n (%) | n (%) |
| Normal | 7 (7.5%) | 11 (11.8%) | 75 (80.6%) |

### Table IV: Preoperative anaesthesia assessment

| Yes | No | Not recorded |
|-----|----|--------------|
| **Bisphosphonate treatment** | n (%) | n (%) | n (%) |
| 73 (78.5%) | 7 (7.5%) | 13 (13.9%) |
| **Airway** | | | |
| Reduced mouth opening | 0 (0%) | 77 (82.7%) | 16 (17.2%) |
| MP 3/4 | 5 (5.4%) | 68 (92.4%) | 20 (21.5%) |
| Decreased neck movement | 0 (0%) | 90 (96.7%) | 3 (3.2%) |
| Spinal deformity | 38 (40.8%) | 27 (29.0%) | 28 (31.1%) |
| Abnormal chest auscultation | 35 (37.6%) | 57 (61.2%) | 1 (1.1%) |
| Abnormal cardiac auscultation | 0 (0%) | 90 (96.7%) | 3 (3.2%) |
| Sedative premedication | 71 (76.3%) | 22 (23.7%) | – |
The intra- and postoperative complications are highlighted in Table VI. Three patients had airway complications; eight patients had difficulty relating to the neuraxial block although only four failed (4.9%); and six patients received a blood transfusion in the perioperative period. All but one patient were treated in a paediatric orthopaedic ward postoperatively. Just over half the patients (55.9%) had a documented rise in temperature intraoperatively. The highest temperature recorded intraoperatively was 38.6°C but from a baseline of 38.1°C. No temperature intraoperatively. The highest temperature recorded were treated in a paediatric orthopaedic ward postoperatively. All but one patient only four failed (4.9%); and six patients received a blood transfusion in the perioperative period. All but one patient were treated in a paediatric orthopaedic ward postoperatively. Just over half the patients (55.9%) had a documented rise in temperature intraoperatively. The highest temperature recorded intraoperatively was 38.6°C but from a baseline of 38.1°C. No children had documented signs suggestive of hypermetabolism or malignant hyperthermia.

| Table VI: Intraoperative and postoperative complications |
|----------------------------------------------------------|
| **Intraoperative complications** | **n (%)** | **N = 81** |
| **Airway** | | |
| Difficult to position SGAD | 3 (3.22%) |
| Difficult to position ETT | 1 |
| Difficult to insert SGAD | 1 |
| **Regional (n = 81)** | | |
| Difficult to perform | 4 |
| Failure | 3 |
| Failed to locate caudal space | 1 |
| **Temperature** | | |
| Not recorded | 33 (35.4%) |
| Decrease | 8 (8.6%) |
| Increase | 52 (55.9%) |
| Increase 0–1°C | 30 |
| Increase 1–3°C | 22 |
| **Blood transfusion** | | |
| Intraoperative | 4 (4.30%) |
| Postoperative | 2 (2.15%) |
| Postop complications | 4 (4.30%) |
| Postop high care | 1 (1.07%) |

**Discussion**

Osteogenesis imperfecta can present many challenges in the perioperative period which include difficulties with airway management, regional techniques and positioning; a risk of bleeding due to platelet dysfunction; respiratory and cardiac compromise; and a risk of hyperthermia. All of these have to be carefully considered before anaesthetising these patients.

Two large case series regarding the anaesthetic management of paediatric OI patients have been published from high income countries; a study published in 1992 by Hall et al. included 266 cases, and a study published in 2018 by Rothschild et al. included 205 cases. Our study is, to our knowledge, the first study describing management of such cases in a low-middle-income country.

All the patients in our study had severe OI and the majority had type III OI. Different genetic populations may account for the different incidence in the severity and type of OI seen in different studies. In our study the majority of our patients were black. The incidence of OI type III was higher in our patients, which was similar to a study done in Australia, where the incidence of OI type III was higher in the black population as compared to the white population, who had a higher incidence of type I OI.

This is in contrast to other studies done on predominately white populations in Sweden13 and Canada,14 where type I OI was found to be the most prevalent type and type III OI was much less common. As expected, and similarly to our surgical cohort, the surgical populations described by Hall et al. and Rothschild et al. had a high incidence of type III OI as type III OI is more severe and often requires more surgery. Furthermore, in our study, only nine patients had a positive family history of OI, which differs from the Swedish cohort, where 87% of their cohort had a positive family history and a causative mutation identified.

In our study we found a very high incidence (65%) of preoperative anaemia. More than half of these were diagnosed based on a finger prick haemoglobin, with no further laboratory testing done. The reason for the anaemia is thus unclear, but could be related to chronic or co-existing disease or recurrent surgical operations. Despite the high incidence of anaemia, few patients (6.5%) required perioperative blood transfusion. This is very different from a study by Hall et al., where more than half of their patients required intraoperative blood transfusion. The Rothschild et al. study showed that 17% of their patients had significant blood loss, mainly patients with type III and IV OI. In our study, pretesting and pretreatment of anaemia by iron and/or folate supplementation was identified as an area of improvement in our future care of patients with OI. Other measures to decrease the rate of blood transfusions could include pretreatment with erythropoietin; the use of tourniquets; and shorter surgical duration. Patients with OI can also have platelet affection secondary to a lower amount of collagen available for platelet aggregation, decreased production of factor VIII and capillary fragility. A preoperative full blood count would allow...

| Table V: Intraoperative anaesthesia conduct |
|--------------------------------------------|
| **Induction** | | |
| Intravenous | 9 (9.6%) |
| Inhalational | 79 (84.9%) |
| Combined | 5 (5.3%) |
| **Airways** | | |
| ETT cuffed | 4 (4.3%) |
| ETT not cuffed | 1 (1.0%) |
| Face mask | 3 (3.3%) |
| SGAD | 85 (91.9%) |
| **Type of anaesthesia** | | |
| Regional + GA | 81 (87.0%) |
| GA alone | 12 (12.9%) |
| **Maintenance** | | |
| Isoflurane | 7 (7.5%) |
| Sevoflurane | 86 (92.4%) |
| **Drugs** | | |
| NMBA | 1 (1.0%) |
| Opioid | 33 (35.4%) |
| NSAID | 73 (78.4%) |
| Ketamine | 24 (25.8%) |
| **Regional technique** | | |
| 81 (87.1%) |
| Neuraxial | 57 (61.2%) |
| Peripheral | 24 (25.8%) |

ETT – endotracheal tube, SGAD – supraglottic airway device, GA – general anaesthesia, NMBA – neuromuscular blockade agent, NSAID – non-steroidal anti-inflammatory drug.
for platelet analysis to pick up an abnormal platelet count and alert the anaesthesiologist to potential bleeding complications.

The main cause of death in OI patients, especially patients with type III OI, is related to respiratory insufficiency secondary to kyphoscoliosis, chest infections and cardiac dysfunction. Respiratory insufficiency can lead to perioperative respiratory complications. Despite the known risk of respiratory disease, and one in three of our patients presenting with abnormal findings on respiratory examination, few patients had pulmonary function testing and/or a chest X-ray (CXR) done preoperatively. Of the 29 patients who had a CXR, 17 had abnormal findings. Only eight patients had pulmonary function testing. Nevertheless, we had no documented cases of intra- or postoperative respiratory complications. Although no details regarding perioperative respiratory complications were given in Hall’s study, they did mention that none of their patients were admitted to ICU postoperatively for ventilation.

Abnormal myocardial collagen can lead to cardiac dysfunction, including mitral valve prolapse, mitral incompetence, aortic root dilatation, aortic incompetence, aortic dissection and ventricular rupture. The degree of cardiac involvement does not seem to correlate with the degree of musculoskeletal involvement. Wong et al. reviewed patients with OI presenting for cardiac surgery. The most common complication in their study was bleeding despite normal coagulation tests, and difficult surgery due to friable tissue and subsequent prosthetic failure. In our study, routine cardiac investigation in the preoperative period was not done and other cardiac involvement could have been missed; however no abnormalities were documented on preoperative examination, and no perioperative cardiac complications were documented. However, because of the risk of cardiac involvement, a thorough preoperative cardiac examination and ECG should be recommended for all patients.

Airway management in patients with OI can be very challenging. Bag-mask ventilation and laryngoscopy may be difficult because of muscle biopsy done two years later was normal, and malignant hyperthermia. The patient had a trigger-free general anaesthesia, but still developed hyperthermia, and the maximum recorded temperature in our cohort was 36.6°C. This was similar to Bojanic’s cohort where their patients had a mild increase in temperature intraoperatively, but was not different to the intraoperative rise in temperature in patients without OI.

In previous studies, children with type III OI had a higher rate of anaesthetic complications. In the Rothschild cohort about half of those patients were admitted to ICU postoperatively for ventilation. A Proseal LMA® may serve as a useful method to insert a temperature monitor and to deflate the stomach. Difficulties with airway management were documented in three patients (3.2%) in our cohort, which included difficulty or inability to insert, or position the SGAD. Our findings are comparable to studies done by Hall et al. and Rothschild et al. who also found a low incidence of difficult airway management. In the study done by Hall et al. most of their patients were managed by SGAD with few difficulties; only two cases had a difficult laryngoscopy. In the study done by Rothschild et al. there were also few documented difficulties (1.5%) with regards to airway management. This relative infrequency of airway complications occurring in our cohort, as well as the other two large cohorts, may be due to anaesthesia provider experience in management of these children.

Regional and neuraxial anaesthesia might be difficult in these patients because of abnormal spinal anatomy. Potential bleeding tendency should also be taken into account when a decision is made to perform regional anaesthesia. The benefits of regional anaesthesia for these patients are mainly due to the opioid-sparing effect. In our study we had a high success rate with regional analgesia which could have decreased the opioid requirement in the perioperative period. Difficulty with RA was experienced in eight cases (9.8%), although only half of these failed. The rate of difficulty is similar to that described by Rothschild (12.5%), who reported most difficulties in patients with OI type III and IV. They had no failures using peripheral nerve blocks although neuraxial failures are not reported.

There is a concern regarding malignant hyperthermia (MH) and hyperpyrexia in children with OI. The association between OI and pyrexia is unclear. The reason for the rise in temperature may be multifactorial and could be related to drugs given as a premedication like hyoscine, use of forced air warmers, an increased thyroxin level, or a hypermetabolic state, rather than MH. Porsborg et al. described a case with OI type III with hypermetabolism in the intraoperative period which was not due to malignant hyperthermia. The patient had a trigger-free general anaesthesia, but still developed hyperthermia, acidosis and tachycardia. Dantrolene was administered, but the temperature and heart rate continued to increase. A muscle biopsy done two years later was normal, and malignant hyperthermia was excluded. Even though there was a documented rise in body temperature in more than half of our cases (55.9%) intraoperatively, we had no cases of malignant hyperthermia, and the maximum recorded temperature in our cohort was 38.6°C. This was similar to Bojanic’s cohort where their patients had a mild increase in temperature intraoperatively, but was not different to the intraoperative rise in temperature in patients without OI.

In our study, most of our patients had their airway secured using a SGAD; only five per cent were intubated, none of which were documented as difficult. SGADs have been shown to be a safe and effective method of airway management in children with OI. According to the literature, SGADs and especially intubating SGADs can be very useful in OI patients as they can prevent airway injury and serve as a rescue device if a difficult laryngoscopy is encountered. A Proseal LMA® may serve as a useful method of securing the airway as the drainage port can also be used
We included all patients with OI that underwent surgery at IALCH between 2000 to 2017. IALCH is a major referral centre for surgical management of patient with OI in the KwaZulu-Natal province; however it is not the only hospital offering surgical management to children with OI. One limitation may be that it was a single centre study, and children with OI operated at other hospitals were not included. Further, all patients were managed by a single orthopaedic team and paediatric anaesthetists, experienced in the management of children with OI.

Another limitation of our study is that it was retrospective. We relied on a chart review and good documentation of the anaesthetic management. We found underreporting of difficult IV access, site of temperature monitoring and blood loss.

**Recommendations**

In our study we had fewer anaesthetic complications than previous studies, despite our higher incidence of type III OI. However, patients with OI should still be managed by a multidisciplinary team in the perioperative period to minimise the risk. The following should be considered:

1. Preoperative assessment is critical for all patients with OI and should include the following:
   - Determine the type and severity of OI, the drug history, previous anaesthetic history and complications.
   - Careful airway and clinical assessment including a cardiac and respiratory exam and excluding abnormalities that may make regional and neuraxial anaesthesia difficult.
   - Investigations to exclude anaemia, bleeding diathesis, electrolyte abnormalities, and cardiac and respiratory investigation if needed.

2. Intraoperative management with emphasis on the following:
   - Careful positioning of the patient to avoid fractures. The use of a tourniquet and a blood pressure cuff can also lead to complications.
   - Temperature monitoring.
   - Monitoring and management of blood loss.
   - Bag-mask ventilation should be done gently to avoid mandibular fractures.
   - SGAD can be used successfully in most patients.
   - If intubation is needed, videolaryngoscopy is advised.
   - Regional anaesthesia is useful to decrease intra- and postoperative opioid requirements and subsequent postoperative respiratory complications.

**Conclusion**

We have demonstrated that the population of surgically managed OI children at IALCH had a high incidence of severe type III OI. Despite this being a high-risk surgical group, most patients were anaesthetised without complication. The majority were managed with a SGAD, and few intraoperative or postoperative complications were documented. However, due to the high incidence of anaemia, improved preoperative investigation, including a careful history and examination and a full blood count, should be encouraged to improve overall perioperative care in patients with OI.

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**Conflict of interest**

The authors declare no conflict of interest.

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