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Management of Severe Scoliosis with Pulmonary Arterial Hypertension: A Single-Center Retrospective Case Series Study

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Abstract

Aims: To determine the impact of anesthesia encountered and to optimize the treatment of perioperative pulmonary arterial hypertension (PAH) in an effort to improve perioperative management and reduce complications. **Methods:** We conducted a retrospective analysis of scoliosis patients with PAH who underwent scoliosis surgery. **Results:** During this period, we identified a total of 22 patients. Their mean age was 22.18 ± 2.11 years. 16 PAH patients (72.72%) received PAH-specific treatment. Only Propofol-based TIVA was used intraoperatively. During the procedure, pulmonary artery catheters and PICCO catheters were placed in all patients to monitor intraoperative and postoperative mPAP, MAP, PRVI and SRVI. During tracheal intubation and intraoperative awake testing, mPAP generally tended to increase in all patients. 6 patients (27.27%) received intraoperative PAH-Specific therapy. All patients received oral sildenafil (75-100 mg/d orally), and 9 patients received postoperative oral sildenafil combined with nebulized iloprost (20 μg/d); intravenous treprostinil (2 ng/kg/min started and titrated to 10-17.5 ng/kg/min); or bosentan (250 mg/d) postoperatively. 7 patients (31.82%) reported postoperative complications, including 2 cases of respiratory failure requiring reintubation, 1 case of right heart failure, 2 cases of superficial surgical site infection, 1 case of fluid and electrolyte and acid-base imbalances, 2 cases of pneumonia and 1 case of pulmonary oedema with fluid overload. Two patients developed more than 1 postoperative complication. No in-hospital death occurred. **Conclusions:** The anesthetic management of scoliosis patients with PAH is important task that, like its own surgery, relies on the input of the multidisciplinary team for its success. Close monitoring, optimization of systemic blood pressure, pain control, oxygenation and ventilation, avoidance of exacerbating factors, and the use of vasopressors and pulmonary vasodilators when necessary are essential elements of management.

Keywords

anesthetic management, pulmonary arterial hypertension, scoliosis surgery

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Introduction

Severe scoliosis has numerous adverse effects on the cardiorespiratory system. Previous studies have shown that scoliosis compromises chest-wall movement and is a causative factor in restrictive ventilatory disorders including reduced forced vital capacity (FVC) as well as a decrease in the percentage of predicted FVC (%FVC). Several early studies indicated that untreated scoliosis increases mortality caused by respiratory failure and right heart failure.[1,2] Pulmonary arterial hypertension (PAH) is thought contribute to the development of cardiac consequences. In scoliosis patients with PAH, anesthesia and scoliosis surgery are associated with a significant increase in morbidity and mortality, mainly due to right ventricular failure, arrhythmias, postoperative hypoxemia, and myocardial ischemia.[3] Preoperative risk assessment and successful management of scoliosis patients with PAH undergoing scoliosis surgery involve an understanding of the pathophysiology of the disease, screening of patients at-risk for PAH, analysis of preoperative and operative risk factors, comprehensive multidisciplinary planning, careful intraoperative management, and early recognition and treatment of postoperative complications.

Here, we report a cases series of scoliosis patients with PAH who successfully underwent scoliosis surgery according to the 2015 European Society of Cardiology (ESC)/European Respiratory Society (ERS) guidelines, in order to provide a reference for the perioperative management of such cases.

Methods

Patient Selection

This was a single-center retrospective study. The institutional review board of the XXX approved this study, and the ethics committee waived the requirements for written informed consent. The medical records of all scoliosis patients with PAH who underwent scoliosis surgery between 2013 and 2020 at the XXX were reviewed retrospectively. All cases were confirmed with pulmonary artery catheterization. Mild pulmonary hypertension was defined as a mean pulmonary artery pressure between 25 and 49 mmHg, and severe pulmonary hypertension was defined a as mean pulmonary artery pressure ≥50 mmHg or sPAP ≥70 mmHg. Pulmonary hypertensive crisis (PHC) was defined as an acute increase in pulmonary arterial pressure (PAP) with an sPAP/systolic arterial pressure ratio >.8 with acute right heart (RV) failure and hypoxemia.

Data Collection

Data extraction included patients' demographics including age, race–ethnicity, comorbidities, New York Heart Association functional state before, during, and after scoliosis surgery, 6-minute walk test, breath-hold test and any other existing co-morbidity. Preoperative pulmonary arterial pressure (PAP)- pulmonary artery catheter (PAC), mean PAP (mPAP)-PAC, pulmonary vascular resistance index (PVRI)-PAC, systemic vascular resistance index (SVRI)-pulse contour cardiac output (PICCO), mean arterial pressure (MAP), cardiac index (CI)-PAC, intraoperative PAH-specific therapy, operative time and postoperative PAH therapy, Post-operative duration of mechanical ventilation (PPDMV), complications, and vasopressor dose were among the parameters investigated.

Statistical Analysis

Statistical analyses were performed using SPSS version 22.0 (IBM, Armonk, New York, USA). Data were presented as the mean ± standard deviations (SD) for normally distributed continuous variables and numbers for categorical variables.

Results

Basic Demographic Characteristics of Scoliosis Patients with Pulmonary Arterial Hypertension Underwent Scoliosis Surgery

From 2013 to 2020, a total of 724 scoliosis patients were treated at our hospital, with 137 cases of idiopathic scoliosis, 212 cases of congenital scoliosis, 114 cases of neuromuscular scoliosis, 129 cases of degenerative scoliosis, 47 cases of traumatic scoliosis, 3 cases of tumour scoliosis, and 82 cases of inflammatory or specific infectious scoliosis. For patients with moderate-to-severe PAH, PAH-specific therapy was initiated before surgery. Most of these patients underwent scoliosis surgery under general anesthesia. A pulmonary artery catheter was placed before the patient was transferred to the operating room.

Twenty-two scoliosis patients were identified with PAH, with an incident rate of 3.04%. Their mean ages were 22.18 ± 2.11 years. Of the 22 patients with combined PAH who underwent scoliosis surgery, 19 patients (86%) had neuromuscular and congenital scoliosis. Their baseline characteristics, and management before scoliosis surgery were listed in Table 1. 13 patients (59.09%) were newly diagnosed with PAH during their hospitalization. 9 patients (40.91%) were previously diagnosed with PAH, and only 2 (9.10%) had received PAH specific therapy. 16 patients (72.73%) had other cardiopulmonary abnormalities, including 14 cases (63.63%) extremely pulmonary dysfunction, 11 (50.00%) structural malformation of heart. 12 (54.55%) had a history of respiratory failure, defined as arterial oxygen tension (PaO2) lower than 60 mmHg with a normal or high arterial carbon dioxide tension (PaCO2). Of these 22 patients, 13 (59.09%) were wheel chair bound. All

Table 1. Demographic Characteristics and Clinical Features.

Parameter	Value
Age (years)	31.6 ± 3.2
Weight (kg)	35.1 ± 12.9
Cobb's angle (o)	127.5 ± 26.8
Height (cm)	126.2 ± 13.8
Arm length (cm)	146.6±19.1
HR (beats/min)	89.2 ± 26.1
BP (mmHg)	123.5 ± 36.5
SpO2 (%)	89.4 ± 7.8
Na^+ (mmol/L)	138.6 ± 2.4
K^+ (mmol/L)	3.9 ± 0.4
Blood urea nitrogen (BUN) (mmol/L)	4.7 ± 1.3
Creatinine (μmol/L)	42.5 ± 6.4
Hb (g/dl)	126.3 ± 11.4
WBC ($\times 10^9$/L)	8.4 ± 1.9
PLT ($\times 10^9$/L)	218.2 ± 31.7

Values are expressed as mean±SD.

patients recovered in the surgical intensive care unit for 3 to 5 days and continued to receive treatment for PAH. Once a patient was hemodynamically stabilized, the patient was transferred to the general ward for additional care.

Preoperative Therapy and Management

16 PAH patients (72.72%) received PAH-specific therapy (oral sildenafil, inhaled iloprost, or treprostinil) after initial diagnosis, 3 to 6 months before scoliosis surgery. They received sildenafil alone or in combination with inhaled iloprost, treprostinil or bosentan (Table 2). All 22 scoliosis patients received traction with a cephalopelvic ring upon admission. 10 scoliosis patients with combined type 2 respiratory failure were on noninvasive-ventilator assisted breathing. All of these patients received oxygen therapy. Only 2 patients received preoperative anticoagulation to prevent thrombosis and digoxin for heart failure. All patients were monitored postoperatively in the intensive care unit and received carefully planned pain management strategies to reduce sympathetic activation and increased pulmonary vascular resistance (PVR), and immediate postoperative monitoring in the intensive care unit. In addition to the above medical measures, all patients were asked to blow up balloons and sing exercises, and for patients with limited mobility, they are also asked to stake the stairs to improve their lung function.

Intraoperative Details of Scoliosis Surgery With Pulmonary Arterial Hypertension

Preoperatively, patients were classified as class 4 according to American Society of Anesthesiologists classification. The majority of procedures were performed in the prone position (81.82%, n = 18), with 4 patients receiving the lateral position due to excessive kyphoscoliosis. After local oral mucosal anesthesia and cricothyroid membrane puncture anesthesia, tracheal intubation was performed. 18 (81.82%) patients were guided by fiberoptic bronchoscope and 4 (18.18%) by visible light wand. Intraoperatively, only propofol-based total intravenous anesthesia (TIVA) was used with various neuromuscular blocking agents (Rocuronium or Cisatracurium), analgesics (Remifentanil, Sufentanil and Oxycodone), and adjuvants (Dexmedetomidine). The median operative time for primary procedures were 386 minutes (IQR 276–469). The estimated total intraoperative blood loss was 1130 mL (IQR 860–2600). 4 patients (18.18%) underwent revisional scoliosis surgery; their median operative time and estimated intraoperative blood loss were 513 minutes (IQR 457-587) and 1684 mL (IQR 1350-2073), respectively (Table 3).

Pulmonary artery catheters were placed in all patients before they were transferred to the operating room. Intraoperative and postoperative monitoring of mPAP, PVR index, systemic vascular resistance (SVR) index, and cardiac output was performed. When the intraoperative mean PAP/mean systemic arterial pressure ratio increased, iloprost was inhaled or intravenous treprostinil was administered to control pulmonary hypertension. Intraoperatively, 2 patients received iloprost inhalation and 5 patients received treprostinil intravenously. Most pulmonary artery catheters were removed within 3 to 5 days after surgery when the patient's condition became relatively stable. The bispectral index (BIS) monitor was used as a guide for anesthetic depth to reduce incidents of sudden hemodynamic disturbances. Somatosensory evoked potentials (SSEPs) and myogenic motor evoked potentials (MMEPs) were monitored in all procedures. During the operation, we also monitored invasive arterial blood pressure and pulse-indicated continuous cardiac output (PICCO). 16 patients experienced intraoperative hypotension (systemic blood pressure < 90/60 mmHg) and were treated with phenylephrine; noradrenaline; or epinephrine to maintain hemodynamic stability. During tracheal intubation and intraoperative awake testing, mPAP generally tended to increase in all patients (Figure.1).

Postoperative Management and Clinic Outcomes

Echocardiography was performed daily after scoliosis surgery. LMWH was started 24 hours after surgery. The pulmonary artery catheter was removed on the third to fifth postoperative day without catheter-related complications. No patient died postoperatively due to pulmonary hypertension and related complications. The mean PAP, PVR index, mean systemic arterial pressure, SVR index, and cardiac index were almost equivalent preoperatively and 72 hours postoperatively (Figure.1). All patients were

Table 2. Baseline Characteristics and Management of Scoliosis Patients with Pulmonary Arterial Hypertension.

Patients	Age (years)	Type of Scoliosis	WHO FC at Hospital Admission	PAH-specific or Other Therapy	SpO2 (%) at Hospital Admission	SPAP-ECHO (mmHg) at Hospital Admission	6MWD at Hospital Admission(m)	NT-proBNP at PAH Diagnosis (ng/L)	PAH Severity	Type 2 Respiratory Failure
1	21	IS	II	None or none	96	74	481	485	Low risk	No
2	24	NS	III	B 125 mg/d, N 20 mg/d or NIV	90	82	248	671	High risk	Yes
3	19	CS	III	None or none	92	76	501	1083	High risk	No
4	23	NS	III	S 75 mg/d or NIV	90	89	420	893	High risk	Yes
5	24	IS	II	S 75 mg/d or none	96	55	486	512	Moderate risk	No
6	26	NS	IV	S 100 mg/d, Ilo 20 μg/d, F 20 mg/d, D 0.25 mg/d, LMWH or NIV	84	98	120	2630	High risk	Yes
7	21	IS	II	B 125 mg/d or none	94	68	508	413		No
8	24	NS	IV	S 100 mg/d, B 125 mg/d, F 20 mg/d, D 0.25 mg/d, LMWH or NIV	86	96	294	1937	High risk	Yes
9	25	NS	IV	S 100 mg/d, N 20 mg/d or NIV	80	113	286	2346	High risk	Yes
10	21	IS	III	None or none	89	63	483	964	High risk	No
11	22	CS	III	S 100 mg/d or none	91	82	462	861	Moderate risk	No
12	22	IS	II	S 75 mg/d or none	92	54	496	635	Moderate risk	No
13	26	NS	III	Ilo 20 μg/d or NIV	88	75	381	1359	High risk	Yes
14	21	CS	II	S 75 mg/d or none	94	72	452	394	Moderate risk	No
15	19	IS	III	None or none	91	94	284	619	Moderate risk	No
16	20	IS	II	None or none	93	48	452	268	Moderate risk	No
17	20	CS	IV	None or NIV	82	93	157	1649	High risk	Yes
18	23	NS	III	S 100 mg/d, Ilo 20 μg/d or NIV	89	64	424	866	High risk	Yes
19	24	CS	III	None or NIV	84	81	392	946	High risk	Yes
20	21	CS	II	S 75 mg/d or none	93	61	436	268	Low risk	No
21	20	IS	I	None or none	95	49	447	123	Low risk	No
22	22	NS	III	S 100 mg/d or NIV	88	68	299	967	High risk	Yes

NS: Neuromuscular Scoliosis; IS: Idiopathic Scoliosis; CS: Congenital Scoliosis; B: Bosentan; N: Nifedipine; NIV: Non-invasive Ventilator therapy; S: Sildenafifil; Ilo: Iloprost; F: Furosemide; D: Digoxin; LMWH: Low-molecular–weight heparin; PHS: Pulmonary Arterial Hypertension.

Table 3. Perioperative Data of Scoliosis Patients with Pulmonary Arterial Hypertension.

Patients	PAP-PAC Pre-operative (mmHg)	Mean PAP-PAC Pre-operative (mmHg)	PVRI-PAC Pre-operative (dynes·s/$cm^5 \cdot m^2$)	SVRI-Picco Pre-operative (dynes·s/cm^5 m^2)	MAP Pre-operative (mmHg)	CI-PAC Pre-operative (L/min·m^2)	Operative Body Position	Intraoperative PAH-specific Therapy	Operative Duration (min)	Intraoperative Blood Loss (ml)
1	62/34	43	384	2238	86	3.1	Prone position	None	324	1050
2	96/71	79	592	1986	72	2.6	Prone position	Ilo 20 μg	486	1680
3	83/60	68	684	2461	80	2.4	Prone position	Ilo 20 μg	596	2608
4	96/45	62	692	1793	71	2.7	Prone position	None	686	1840
5	74/34	47	319	1933	92	3.4	Prone position	None	351	1294
6	112/68	83	1011	2468	106	2.0	Lateral position	Tre 2 ng/kg/min	468	1845
7	52/34	40	336	2018	81	3.1	Prone position	None	513	860
8	110/63	79	916	1930	96	1.8	Lateral position	Tre 2 ng/kg/min	512	2684
9	86/45	59	715	2096	84	2.2	Prone position	Tre 2 ng/kg/min	608	1682
10	76/40	52	496	2461	90	2.7	Prone position	None	691	2643
11	94/48	63	618	2715	106	2.4	Prone position	None	705	1620
12	61/48	52	351	2184	89	3.1	Prone position	None	439	2662
13	82/61	68	634	2291	96	2.6	Prone position	None	563	1686
14	69/54	59	412	2683	105	3.2	Prone position	None	324	1832
15	90/42	58	643	1925	76	2.6	Prone position	None	584	1680
16	61/54	56	346	1897	81	3.8	Prone position	None	431	2086
17	98/47	64	851	2843	103	1.9	Lateral position	Tre 2 ng/kg/min	572	1368

(continued)

Table 3. (continued)

Patients	PAP-PAC Pre-operative (mmHg)	Mean PAP-PAC Pre-operative (mmHg)	PVRI-PAC Pre-operative (dynes·s/ $cm^5 \cdot m^2$)	SVRI-Picco Pre-operative (dynes·s/ cm^5 m^2)	MAP Pre-operative (mmHg)	CI-PAC Pre-operative (L/min·m^2)	Operative Body Position	Intraoperative PAH-specific Therapy	Operative Duration (min)	Intraoperative Blood Loss (ml)
18	92/40	57	761	2034	96	2.7	Prone position	None	496	968
19	106/60	75	944	3012	112	2.4	Lateral position	None	536	1094
20	58/39	45	346	2433	102	3.6	Prone position	None	463	1389
21	55/38	44	358	2196	92	3.6	Prone position	None	582	2068
22	82/66	71	539	2681	107	2.8	Prone position	None	468	1968

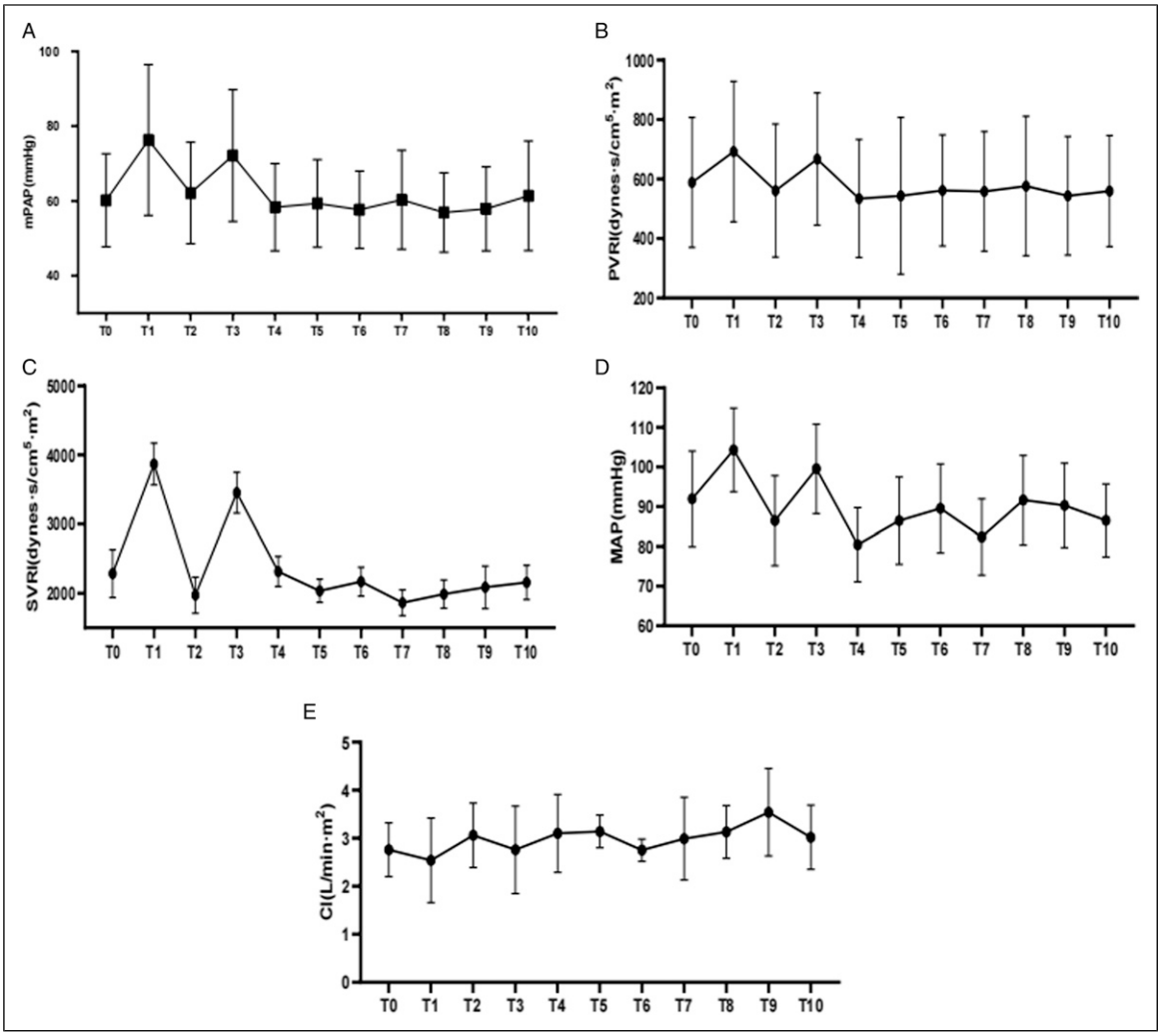

Figure 1. Perioperative hemodynamic parameters over 72 hours for mPAP (A), PVRI (B), SVTI (C), Map (D) and CI(E). mPAP: mean systemic pulmonary arterial pressure; PVRI: pulmonary vascular resistance index; SVRI: systemic vascular resistance index; MAP: mean systemic arterial pressure; CI: cardiac index. The horizontal axis of the coordinates indicatingT0, at induction; T1, at intubation; T2, at skin incision; T3, at intraoperative awake-test; T4, at the end of the procedure; T5, 12 hours postoperatively; T6, 24 hours postoperatively; T7, 36 hours postoperatively; T8, 48 hours postoperatively; T9, 60 hours postoperatively; T10, 72 hours postoperatively.

treated with oral sildenafil (75-100 mg/d orally), and 9 received oral sildenafil combined with nebulized iloprost (20 μg/d); intravenous treprostinil (starting at 2 ng/kg/min and titrated to 10-17.5 ng/kg/min); or bosentan (250 mg/d) based on PAP measurements by pulmonary artery catheter (Table 4).

Early postoperative complications (≤30 d) were reported in 7 patients (31.82%). Major early complications included respiratory failure requiring reintubation (n = 2), right heart failure (n = 1). Minor early complications included superficial surgical site infection (n = 2), fluid and electrolyte and acid-base imbalances (n = 1), pneumonia (n = 2), pulmonary edema with fluid overload (n = 1). Two patients developed >1 early postoperative complications. Overall, 4 patients developed cardiopulmonary complications. All patients received post-operative analgesia, including oral NSAIDs and intravenous oxycodone. There were no postoperative deaths in this cohort.

Discussion

PAH is a known risk factor for perioperative complications. Most of the literature regarding the role of PAH in perioperative morbidity and mortality in patients undergoing noncardiac surgery is based on small series and remains poorly defined.[4] PAH is currently not listed in the American Heart Association/American College of Cardiology Foundation Practice Guideline for noncardiac

Table 4. Postoperative Management of Scoliosis Patients with Pulmonary Arterial Hypertension.

Patients	Post-operative Pulmonary Arterial Hypertension Therapies	Other Therapies	PPDMV (h)	Post-operative Complications	Dosage of Vasopressor
1	S 75 mg/d, Ilo 20 μg/d, LMWH 5000IU/d	None	3	None	Norepinephrine .03 mg/kg/min
2	B 125 mg/d, N 20 mg/d, LMWH 5000IU/d	Non-invasive ventilator therapy	11	None	Norepinephrine .05 mg/kg/min
3	S 100 mg/d, Ilo 20 μg/d, LMWH 5000IU/d	CRRT	16	FEAI	Epinephrine .02 mg/kg/min
4	S 75 mg/d, LMWH 5000IU/d	Non-invasive ventilator therapy	37	Pulmonary edema	Norepinephrine .05 mg/kg/min
5	S 75 mg/d, Ilo 20 μg/d, LMWH 5000IU/d	None	4	None	None
6	S 100 mg/d, Ilo 20 μg/d, furosemide 20 mg/d,digoxin,0.25 mg/d, LMWH 5000 IU/d	Non-invasive ventilator therapy	61	Respiratory failure, pneumonia	Epinephrine .02 mg/kg/min
7	Bosentan 125 mg/d, LMWH 5000IU/d	None	11	None	Norepinephrine .01 mg/kg/min
8	S 100 mg/d, bosentan 125 mg/d, furosemide 20 mg/d, digoxin,0.25 mg/d, LMWH 5000 IU/d	Non-invasive ventilator therapy	72	Respiratory failure, pneumonia	Epinephrine .01 mg/kg/min, dobutamine 5 μg/kg/min
9	S 100 mg/d, Nifedipine 20 mg/d, LMWH 5000IU/d	Non-invasive ventilator therapy	28	None	Norepinephrine .05 mg/kg/min, dobutamine 5 μg/kg/min
10	S 100 mg/d, Nifedipine 20 mg/d, LMWH 5000IU/d	None	16	None	Norepinephrine .03 mg/kg/min
11	S 100 mg/d, Ilo 20 μg/d, LMWH 5000IU/d	None	12	Surgical site infection	Norepinephrine .04 mg/kg/min
12	S 75 mg/d, Ilo 20 μg/d, LMWH 5000IU/d	None	9	None	None
13	Ilo 20ug/d, LMWH 5000IU/d	Non-invasive ventilator therapy	24	None	Norepinephrine .04 mg/kg/min
14	S 75 mg/d, LMWH 5000IU/d	None	16	Surgical site infection	None
15	S 75 mg/d, LMWH 5000IU/d	None	33	None	Epinephrine .01 mg/kg/min
16	S 75 mg/d, LMWH 5000IU/d	None	26	None	None
17	S 100 mg/d, Nifedipine 20 mg/d, furosemide 20 mg/d, digoxin,0.25 mg/d, LMWH	Non-invasive ventilator therapy	63	Right heart failure	Epinephrine .03 mg/kg/min and dobutamine 5 μg/kg/min
18	S 100 mg/d, Ilo 20 ug/d, LMWH 5000IU/d	Non-invasive ventilator therapy	37	None	Norepinephrine .05 mg/kg/min
19	S 100 mg/d, LMWH 5000IU/d	Non-invasive ventilator therapy	28	None	Norepinephrine .05 mg/kg/min
20	S 75 mg/d, Ilo 20 ug/d, LMWH 5000IU/d	None	19	None	None
21	S 75 mg/d, LMWH 5000IU/d	None	8	None	None
22	S 100 mg/d, Ilo 20 μg/d, LMWH 5000IU/d	Non-invasive ventilator therapy	42	None	None

PPDMV: Post-operative duration of mechanical ventilation; FEAI: Fluid and electrolyte and acid-base imbalances.

surgery as an independent risk factor for postoperative complications.[5] Despite the paucity of data, patients with PAH are often counseled against having elective procedures because early and sudden postoperative deaths have been reported.[6]

Potential complications, including hypotension, respiratory compromise, or RV failure, may occur intraoperatively or in the postoperative period.[7] Despite the limited literature, several factors have been identified as potential predictors of increased risk of perioperative complications in patients with PAH undergoing noncardiac surgery. Patients with PAH are unable to accommodate alterations in right ventricular (RV) preload or afterload induced by fluid shifts, medications, or changes in the autonomic nervous system precipitated by hypoxia or hypercapnia.[8] These factors become magnified in situations of added stress such as surgical intervention. Systemic hypotension and arrhythmias may precipitate RV ischemia, further worsening RV function.[9,10] Patient and surgical characteristics and choice of anesthetic technique are crucial factors in perioperative management. The 2 main principles of perioperative management are the prevention of systemic hypotension (risk of RV ischemia) and the prevention of acute elevations in pulmonary arterial pressure (risk of RV failure).[11-13] Close monitoring, optimization of systemic blood pressure (BP), pain control, oxygenation and ventilation, avoidance of exacerbating factors, and use of vasopressors and pulmonary vasodilators as necessary are essential elements of management.[8,12,14] Understanding the pathophysiology, cause, and severity of PAH in the individual perioperative patient allows accurate risk assessment, optimization of PAH and RV function prior to surgery, and appropriate intraoperative and postoperative management.[2,15,16]

Appropriate preoperative evaluation and medical optimization, as well as close monitoring during the perioperative period to avoid noxious stimuli, are essential for success. Perioperative medical management of these complex patients often requires a multidisciplinary approach with input from anesthesiologists, PAH experts, pharmacists, and surgeons. In our clinical practice, perioperative management was based on the literature and the expertise of the authors in managing these patients, as there are no evidence-based guidelines.[17,18] Based on the evidence, the patients with PAH undergoing scoliosis surgery are also at increased risk and be evaluated for the cause and severity of the PAH prior to surgery. In addition to the assessment of external scoliosis, we performed a thorough history and physical examination, ECG, chest radiograph and Doppler echocardiography to assess the structure and function of the RV and LV and valves. As an anesthesiologist, several key issues should be considered in the preoperative evaluation, including the risk-benefit ratio of the surgery, the anesthetic technique to be used, the potential for the surgery to disrupt fluid balance or increase PVR, and the availability of necessary tools to prevent and treat PAH and acute right heart failure should they occur. The principles of management include the avoidance of systemic hypotension (risk of RV ischemia), myocardial depression (risk of RV failure due to diminished contractility), acute elevations of PAP (risk of RV failure due to increased impedance), inadequate pain control (risk of increased sympathetic tone and PVR), and respiratory depression.[19-21]

Specific medications being used to treat PAH, such as oral sildenafil, inhaled iloprost, or treprostinil therapy must be continued, as discontinuation induces aa PAH crisis.[8,11,22] Even though some of these medications can inhibit platelet aggregation, this is usually a minor effect and excessive surgical bleeding has not been reported. Throughout the scoliosis surgery procedure, we maintained oxygenation well and avoided systemic hypotension, which could have led right heart ischemia. The standard American Society of Anesthesiologists monitors was used in all scoliosis patients. In addition, we applied an indwelling arterial catheter to monitor the systemic BP during induction of general anesthesia. We also monitored central venous pressure to assess changes in RV filling pressure and to detect new or worsening of tricuspid insufficiency. Since PA catheter monitoring allows accurate assessment of PAP for proper treatment with pulmonary vasodilators, measurement of CO and mixed venous oxygen saturation, as well as accurate measurement of central venous pressure and pulmonary artery pressure. Because of the ability to accurately assess PAP for appropriate treatment with pulmonary vasodilators, measure CO, mixed venous oxygen saturation, calculate PVR, and accurately measure central venous pressure and pulmonary wedge pressure to determine the need for vasopressors and fluids, all of our patients with scoliosis combined with PAH were monitored with PA catheter. We did not perform intraoperative TEE monitoring due to the position restrictions of scoliosis surgery.

All scoliosis patients underwent general anesthesia. The choice of anesthetic technique and anesthetic management are crucial factors in maintaining cardiovascular stability in patients with PAH. However, regardless of the anesthetic technique used, an adequate level of anesthesia should be maintained to produce effective pain control. Good oral mucosal and tracheal surface anesthesia is particularly important because the patient's neck is held in place by an external fixation brace (cephalopelvic ring), the patient's neck is difficult to tilt posteriorly, and all patients were intubated while awake. All the intubation were performed in a rapid and smooth manner by an experienced person. Throughout conscious tracheal intubation, we preoxygenated with 100% oxygen to achieve an end-tidal oxygen concentration of more than 90%, which mitigates the

increased risk of hypoxemia that often occurs after induction due to reduced functional residual capacity. Patients were also instructed to breathe deeply to avoid an increase in arterial CO 2 and to achieve adequate oxygenation. Anesthetics may depress myocardial contractility, decrease SVR and venous return, increase PVR, or exacerbate hypoxia or hypercapnia. Etomidate has the least effect on myocardial contractility and SVR, and in combination with opioids is usually our induction agent of choice. IV or nebulized milrinone or epoprostenol, IV nitroglycerin, inhaled nitric oxide, or nebulized epoprostenol or iloprost can all be used before induction to prevent pulmonary hypertensive responses, although their efficacy has not been studied adequately and selection depends largely on whether they are used preoperatively. When titrating anesthetics, it should be remembered that the metabolism of drugs may be altered in patients with chronic severe PAH due to prolonged venous congestion and visceral dysfunction. During maintenance, we avoid exacerbating risk factors that exacerbate impaired RV, such as hypothermia, hypoxia, hypercarbia, acidosis, and hyper- or hypovolemia. To achieve these goals, blood gases, temperature and ventilation need to be monitored and adequate depth of anesthesia needs to be maintained. A lung-preserving ventilator strategies using low tidal volumes may help avoid overinflation of the lungs and minimize PVR by keeping lung volume close to functional residual capacity. Scoliosis surgery carries greater risks for patients with PAH because of the long operative time, volume shifts and blood loss, and the impact on cardiovascular or pulmonary function. In particular, patients with combined thoracic deformities and intraoperative thoracoplasty may experience a temporary or permanent increase in PVR.

In addition, scoliosis correction may increase intra-abdominal pressure enough to impair pulmonary compliance or reduce venous return. Postoperatively, patients are at risk of worsening PAH values and RV ischemia as the effects of the anesthetics used during surgery, as well as opioids, wear off. Pain increases PVR, but opioids used systemically can have the same effect, as they tend to cause respiratory acidosis. In this case, regional blocks may be beneficial as they relieve pain without inhibiting respiratory drive. We also routinely use adjuvant therapy with NSAIDs intraoperatively to improve pain control.

A feature of the intraoperative management of scoliosis surgery is that in order to determine the function of the spinal cord, the surgeon will require the anesthesiologist to carry out a wake-up test after the main steps of the operation have been completed. In scoliosis patients with combined PAH, this is a huge risk. This is why we gave an adequate amount of analgesic medications, including oxycodone or NSIADs during intraoperative arousal in these patients. Arousal times are usually longer than in other patients with scoliosis without PAH. Despite this, during our PA catheter monitoring, we found that the patient's pulmonary artery pressure was elevated during intraoperative arousal.

Recommendation

For all patients with scoliosis, we recommend Trans-Thoracic Echocardiography (TTE) prior to admission, which provides a clearer picture of the structure of the heart chambers, changes in valve motion and blood flow spectrum, and indirectly infers pressure changes in the pulmonary circulation. When TTE shows a combination of PAH, patients should undergo right heart catheterization upon admission to accurately measure pulmonary artery pressure, which is the "gold standard" for evaluating the accuracy of various non-invasive manometric methods. In addition to accurately measuring pulmonary artery pressure, it can be used to determine pulmonary artery wedge pressure and to measure the oxygen content of the heart chambers, as well as to assess the degree of hemodynamic compromise and to test pulmonary vascular reactivity.

Conclusions

PAH is a high-risk factor for scoliosis surgeries. Careful preoperative evaluation should be performed in all patients with PAH, and those deemed at higher risk by virtue of significant reductions in functional capacity or abnormalities on echocardiogram should undergo a preoperative right-sided heart catheterization to better characterize their severity. Careful planning among PAH specialists, anesthesiologists, and surgeons can help avoid unnecessary complications. Multiple pharmacotherapies, including specific PAH drugs, vasopressors, and orthomimetics, have been used to treat PAH patients in the perioperative period, but have not been systematically studied. On the other hand, observation of fundamental management principles such as optimization of fluid volume, systemic BP, and acid base balance; avoidance of hypoxemia and hypercapnia; and good pain control are clearly important to achieve the desired surgical outcomes.

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References

1. Koumbourlis AC. Scoliosis and the respiratory system. *Paediatr Respir Rev*. 2006;7(2):152-160.
2. Lin Y, Shen J, Chen L, et al. Cardiopulmonary function in patients with congenital scoliosis: An observational study. *J Bone Joint Surg Am*. 2019;101(12):1109-1118.
3. Lin Y, Tan H, Rong T, et al. Impact of thoracic cage dimension and geometry on cardiopulmonary function in patients with congenital scoliosis: A prospective study. *Spine*. 2019;44(20):1441-1448.
4. Aguirre MA, Lynch I, Hardman B. Perioperative management of pulmonary hypertension and right ventricular failure during noncardiac surgery. *Adv Anesth*. 2018;36(1):201-230.
5. Halperin JL, Levine GN, Al-Khatib SM, et al. Further evolution of the ACC/AHA clinical practice guideline recommendation classification system: A report of the American college of cardiology/American heart association task force on clinical practice guidelines. *J Am Coll Cardiol*. 2016;133(14):1426-1428.
6. Schisler T, Marquez JM, Hilmi I, et al. *Pulmonary Hypertensive Crisis on Induction of Anesthesia: Recommendations for Safe Practice; Proceedings of the seminars in cardiothoracic and vascular Anesthesia, F*; Los Angeles, CA: SAGE Publications Sage CA; 2017.
7. Soroceanu A, Burton DC, Oren JH, et al. Medical complications after adult spinal deformity surgery: Incidence, risk factors, and clinical impact. *Spine*. 2016;41(22):1718-1723.
8. Gupta B, Kakkar K, Gupta L, et al. *Anesthetic Considerations for a Parturient with Pulmonary Hypertension; Proceedings of the Indian Anaesthetists Forum, F*. Mumbai, India: Medknow Publications; 2017.
9. Maguire K. *Adolescent Idiopathic Scoliosis Patients are at Increased Risk for Pulmonary Hypertension Which Reverses after Corrective Scoliosis Surgery; Proceedings of the 2014 AAP National Conference and Exhibition, F*. Itasca, IL: American Academy of Pediatrics; 2014.
10. Sarwahi V, Borlack RE, Dworkin A, et al. Adolescent idiopathic scoliosis patients are at increased risk for pulmonary hypertension which reverses after scoliosis surgery. *Spine J*. 2014;14(11):S153-S154.
11. Przybylski R, Hedequist DJ, Nasr VG, et al. Adverse perioperative events in children with complex congenital heart disease undergoing operative scoliosis repair in the contemporary era. *Pediatr Cardiol*. 2019;40(7):1468-1475.
12. Helal SM, Abd Elaziz AA, Dawoud AG. Anesthetic considerations during intraoperative neurophysiological monitoring in spine surgery. *Menoufia Med J*. 2018;31(4):1187.
13. Tellermann J, Sablinskis M, Machado PRR, et al. Long-term response to vasoactive treatment in a case of kyphoscoliosis-associated pulmonary hypertension. *Am J Case Rep*. 2019;20:1505-1508.
14. Bersot CD, Guimarães JE, Mercante R, et al. *Intraoperative Neurophysiologic Monitoring in Spinal Cord Surgery: A Brief Review*. https://www.researchgate.net/publication/348154686_. January 2, 2021.
15. Smilowitz NR, Armanious A, Bangalore S, et al. Cardiovascular outcomes of patients with pulmonary hypertension undergoing noncardiac surgery. *Am J Cardiol*. 2019;123(9):1532-1537.
16. Steppan J, Diaz-Rodriguez N, Barodka VM, et al. Focused review of perioperative care of patients with pulmonary hypertension and proposal of a perioperative pathway. *Cureus*. 2018;10(1):e2072.
17. Inao T, Amano M, Hashimoto S, et al. Rapid improvement of severe pulmonary hypertension due to scoliosis-related restrictive ventilatory disorder. *Intern Med*. 2021:60(20):3289-3293.
18. Seo JS, So KY, Kim SH. Perioperative anesthetic considerations in patients with pulmonary hypertension undergoing non-cardiac and non-obstetric surgeries. *Med Biol Sci Eng*. 2019;2(2):31-39.
19. Gruenbaum BF, Gruenbaum SE. Neurophysiological monitoring during neurosurgery: Anesthetic considerations based on outcome evidence. *Curr Opin Anaesthesiol*. 2019;32(5):580-584.
20. De la Garza Ramos R, Goodwin CR, Abu-Bonsrah N, et al. Patient and operative factors associated with complications following adolescent idiopathic scoliosis surgery: An analysis of 36,335 patients from the nationwide inpatient sample. *J Neurosurg Pediatr*. 2016;18(6):730-736.
21. Bronheim R, Khan S, Carter E, Sandhaus RA, Raggio C: Scoliosis and cardiopulmonary outcomes in osteogenesis imperfecta patients. *Spine*. 2019, 44(15):1057-1063.
22. Latham GJ, Yung D. Current understanding and perioperative management of pediatric pulmonary hypertension. *Paediatr Anaesth*. 2019;29(5):441-456.