

World Journal of *Clinical Cases*

World J Clin Cases 2023 June 16; 11(17): 3932-4209



REVIEW

- 3932 Liver replacement therapy with extracorporeal blood purification techniques current knowledge and future directions

Papamichalis P, Oikonomou KG, Valsamaki A, Xanthoudaki M, Katsiafylloudis P, Papapostolou E, Skoura AL, Papamichalis M, Karvouniaris M, Koutras A, Vaitis E, Sarchosi S, Papadogoulas A, Papadopoulos D

MINIREVIEWS

- 3949 Prediction models for recurrence in patients with small bowel bleeding
Kim JH, Nam SJ
- 3958 Investigation of possible relationship between atopic dermatitis and salivary biomarkers, stress, and sleep disorders
Estefan J, Ferreira DC, Cavalcante FS, dos Santos KRN, Ribeiro M
- 3967 Value of clinical applications of differential pressure and relative pressure imaging in the left ventricle
Zheng AS, Yu HX
- 3976 Low-dose immunotherapy as a potentiator to increase the response with neo-adjuvant chemotherapy in oral cancers
Rathinasamy N, Muthu S, Krishnan A
- 3980 Kidney disease in patients with chronic liver disease: Does sex matter?
Cooper KM, Colletta A, Moulton K, Ralto KM, Devuni D

ORIGINAL ARTICLE

Case Control Study

- 3993 Elabela is a reliable biomarker for predicting early onset preeclampsia: A comparative study
Amer Ali E, Nori W, Salman AF, Al-Rawi TSS, Hameed BH, Al-Ani RM

Retrospective Cohort Study

- 4003 Acute-on-chronic liver failure is independently associated with higher mortality for cirrhotic patients with acute esophageal variceal hemorrhage: Retrospective cohort study
Terres AZ, Balbinot RS, Muscope ALF, Longen ML, Schena B, Cini BT, Rost Jr GL, Balensiefer JIL, Eberhardt LZ, Balbinot RA, Balbinot SS, Soldara J

Retrospective Study

- 4019 Elastic fiber degradation in the development of pediatric granuloma annulare: Report of 39 cases
Zhang DY, Zhang L, Yang QY, Xie YC, Jiang HC, Li JZ, Shu H

- 4026** Anti-bacterial mechanism of baicalin-tobramycin combination on carbapenem-resistant *Pseudomonas aeruginosa*

Jin LM, Shen H, Che XY, Jin Y, Yuan CM, Zhang NH

SYSTEMATIC REVIEWS

- 4035** Acknowledging the use of botanicals to treat diabetic foot ulcer during the 21st century: A systematic review

Narzary I, Swarnakar A, Kalita M, Middha SK, Usha T, Babu D, Mochahary B, Brahma S, Basumatary J, Goyal AK

CASE REPORT

- 4060** Pregabalin induced balance disorder, asthenia, edema, and constipation in an elderly adult: A case report

Ma LP, Wen C, Zhao TX, Jiang XM, Gu J

- 4065** Emergency internal iliac artery temporary occlusion after massive hemorrhage during surgery of cesarean scar pregnancy: A case report

Xie JP, Chen LL, Lv W, Li W, Fang H, Zhu G

- 4072** Hemophagocytic lymphohistiocytosis after autologous stem cell transplantation in angioimmunoblastic T-cell lymphoma: A case report

Zhang ZR, Dou AX, Liu Y, Zhu HB, Jia HP, Kong QH, Sun LK, Qin AQ

- 4079** Successful reconstruction of an ankle defect with free tissue transfer in a hemophilia A patient with repetitive hemoarthrosis: A case report

Lee DY, Lim S, Eo S, Yoon JS

- 4084** Primary pelvic *Echinococcus granulosus* infection: A case report

Abulaiti Y, Kadi A, Tayier B, Tuergan T, Shalayiadang P, Abulizi A, Ahan A

- 4090** Epstein-Barr virus-induced infection-associated hemophagocytic lymphohistiocytosis with acute liver injury: A case report

Sun FY, Ouyang BQ, Li XX, Zhang T, Feng WT, Han YG

- 4098** Cardiac arrest secondary to pulmonary embolism treated with extracorporeal cardiopulmonary resuscitation: Six case reports

Qiu MS, Deng YJ, Yang X, Shao HQ

- 4105** Flared inflammatory episode transforms advanced myelodysplastic syndrome into aplastic pancytopenia: A case report and literature review

Ju B, Xiu NN, Xu J, Yang XD, Sun XY, Zhao XC

- 4117** Frontal penetrating arrow injury: A case report

Rodríguez-Ramos A, Zapata-Castilleja CA, Treviño-González JL, Palacios-Saucedo GC, Sánchez-Cortés RG, Hinojosa-Amaya LG, Nieto-Sanjuanero A, de la O-Cavazos M

- 4123** Chest wall osteochondroma resection with biologic acellular bovine dermal mesh reconstruction in pediatric hereditary multiple exostoses: A case report and review of literature

Alshehri A

- 4133** Massive pulmonary embolism in Klippel-Trenaunay syndrome after leg raising: A case report
Lo CY, Chen KB, Chen LK, Chiou CS
- 4142** Improved super-elastic Ti-Ni alloy wire intrusion arch for skeletal class II malocclusion combined with deep overbite: A case report
Yang CY, Lin CC, Wang LJ, Chen YH, Yu JH
- 4152** Glucocorticoid pulse therapy in an elderly patient with post-COVID-19 organizing pneumonia: A case report
Park S, Jang Y, Koo SM, Nam BD, Yoon HY
- 4159** Endoscopic and surgical treatment of jejunal gallstone ileus caused by cholecystoduodenal fistula: A case report
Fan WJ, Liu M, Feng XX
- 4168** Application of advanced platelet-rich fibrin for through-and-through bony defect during endodontic surgery: Three case reports and review of the literature
Algahtani FN, Almohareb R, Aljamie M, Alkhunaini N, ALHarthi SS, Barakat R
- 4179** Facial Merkel cell carcinoma in a patient with diabetes and hepatitis B: A case report
Ren MY, Shi YJ, Lu W, Fan SS, Tao XH, Ding Y
- 4187** Pregnancy and lactation-associated osteoporosis with pyogenic spondylitis: A case report
Zhai K, Wang L, Wu AF, Qian Y, Huang WM
- 4194** Hourglass-like constriction of the anterior interosseous nerve in the left forearm: A case report
He R, Yu JL, Jin HL, Ng L, Wang JC, Li X, Gai TT, Zhou Y, Li DP
- 4202** Crohn's disease in human immunodeficiency virus-infected patient: A case report
Vinikaite A, Kurlinkus B, Jasinskaite D, Strainiene S, Buineviciute A, Sadauskaite G, Kiudelis V, Kazenaite E

ABOUT COVER

Editorial Board Member of *World Journal of Clinical Cases*, Chun-Lin Ou, Doctor, PhD, Associate Professor, Associate Research Scientist, Department of Pathology, Xiangya Hospital, Central South University, Xiangya Hospital, Central South University, Changsha 410008, Hunan Province, China. ouchunlin@csu.edu.cn

AIMS AND SCOPE

The primary aim of *World Journal of Clinical Cases* (WJCC, *World J Clin Cases*) is to provide scholars and readers from various fields of clinical medicine with a platform to publish high-quality clinical research articles and communicate their research findings online.

WJCC mainly publishes articles reporting research results and findings obtained in the field of clinical medicine and covering a wide range of topics, including case control studies, retrospective cohort studies, retrospective studies, clinical trials studies, observational studies, prospective studies, randomized controlled trials, randomized clinical trials, systematic reviews, meta-analysis, and case reports.

INDEXING/ABSTRACTING

The WJCC is now abstracted and indexed in Science Citation Index Expanded (SCIE, also known as SciSearch®), Journal Citation Reports/Science Edition, Current Contents®/Clinical Medicine, PubMed, PubMed Central, Scopus, Reference Citation Analysis, China National Knowledge Infrastructure, China Science and Technology Journal Database, and Superstar Journals Database. The 2022 Edition of Journal Citation Reports® cites the 2021 impact factor (IF) for WJCC as 1.534; IF without journal self cites: 1.491; 5-year IF: 1.599; Journal Citation Indicator: 0.28; Ranking: 135 among 172 journals in medicine, general and internal; and Quartile category: Q4. The WJCC's CiteScore for 2021 is 1.2 and Scopus CiteScore rank 2021: General Medicine is 443/826.

RESPONSIBLE EDITORS FOR THIS ISSUE

Production Editor: Hua-Ge Yin; Production Department Director: Xiang Li; Editorial Office Director: Jin-Lei Wang.

NAME OF JOURNAL

World Journal of Clinical Cases

ISSN

ISSN 2307-8960 (online)

LAUNCH DATE

April 16, 2013

FREQUENCY

Thrice Monthly

EDITORS-IN-CHIEF

Bao-Gan Peng, Jerzy Tadeusz Chudek, George Kontogeorgos, Maurizio Serati, Ja Hyeon Ku

EDITORIAL BOARD MEMBERS

<https://www.wjgnet.com/2307-8960/editorialboard.htm>

PUBLICATION DATE

June 16, 2023

COPYRIGHT

© 2023 Baishideng Publishing Group Inc

INSTRUCTIONS TO AUTHORS

<https://www.wjgnet.com/bpg/gerinfo/204>

GUIDELINES FOR ETHICS DOCUMENTS

<https://www.wjgnet.com/bpg/GerInfo/287>

GUIDELINES FOR NON-NATIVE SPEAKERS OF ENGLISH

<https://www.wjgnet.com/bpg/gerinfo/240>

PUBLICATION ETHICS

<https://www.wjgnet.com/bpg/GerInfo/288>

PUBLICATION MISCONDUCT

<https://www.wjgnet.com/bpg/gerinfo/208>

ARTICLE PROCESSING CHARGE

<https://www.wjgnet.com/bpg/gerinfo/242>

STEPS FOR SUBMITTING MANUSCRIPTS

<https://www.wjgnet.com/bpg/GerInfo/239>

ONLINE SUBMISSION

<https://www.f6publishing.com>



Massive pulmonary embolism in Klippel-Trenaunay syndrome after leg raising: A case report

Chih-Yu Lo, Kuen-Bao Chen, Li-Kuei Chen, Chiuan-Shiou Chiou

Specialty type: Anesthesiology

Provenance and peer review:

Unsolicited article; Externally peer reviewed.

Peer-review model: Single blind

Peer-review report's scientific quality classification

Grade A (Excellent): 0

Grade B (Very good): B

Grade C (Good): C

Grade D (Fair): 0

Grade E (Poor): 0

P-Reviewer: DeSousa K, India;
Viswanathan VK, United States

Received: March 12, 2023

Peer-review started: March 12, 2023

First decision: April 20, 2023

Revised: April 27, 2023

Accepted: May 12, 2023

Article in press: May 12, 2023

Published online: June 16, 2023



Chih-Yu Lo, Li-Kuei Chen, Chiuan-Shiou Chiou, Department of Anesthesiology, China Medical University Hospital, Taichung 40447, Taiwan

Kuen-Bao Chen, Department of Anesthesiology Pain Service and Critical Care Medicine, China Medical University Hospital, Taichung 40447, Taiwan

Corresponding author: Chiuan-Shiou Chiou, MD, PhD, Doctor, Department of Anesthesiology, China Medical University Hospital, No. 2 Yude Road, Taichung 40447, Taiwan.
chiou.cs@gmail.com

Abstract

BACKGROUND

Klippel-Trenaunay syndrome (KTS) is a rare congenital disorder characterized by a combination of capillary malformations, soft-tissue or bone hypertrophy, and varicose veins or venous malformations. The syndrome predisposes patients to hypercoagulable states, including venous thromboembolism and pulmonary embolism (PE).

CASE SUMMARY

A 12-year-old girl with KTS was scheduled excision of verrucous hyperkeratosis in the left foot and posterior aspect of the left leg and left thigh and excision of a cutaneous hemangioma in the right buttock. After induction, the surgeon elevated the patient's leg for sterilization, whereupon she experienced a massive PE and refractory cardiac arrest. Extracorporeal membrane oxygenation (ECMO) was performed after prolonged resuscitation, and she had a return of spontaneous circulation. After this episode, the patient was discharged without any neurologic complications.

CONCLUSION

The mechanism of PE, a lethal disease, involves a preexisting deep vein thrombosis that is mechanically dislodged by compression or changing positions and travels to the pulmonary artery. Therefore, patients predisposed to PE should be prescribed prophylactic anticoagulants. If the patient has unstable vital signs, resuscitation should be started immediately, and extracorporeal cardiopulmonary resuscitation should be considered in settings with existing ECMO protocols, expertise, and equipment. Awareness of PE in patients with KTS while leg raising for sterilization is critical.

Key Words: Klippel-Trenaunay syndrome; Pediatric; Pulmonary embolism; Anesthesia; Case report

©The Author(s) 2023. Published by Baishideng Publishing Group Inc. All rights reserved.

Core Tip: Klippel-Trenaunay syndrome (KTS) is a congenital vascular disorder. Patients with KTS are at high risk for pulmonary embolism. All patient having KTS should be evaluated of lower limb circulation and pre-existing deep vein thrombosis (DVT) pre-operatively. Also, all patients with KTS should receive DVT prophylaxis at least 8 hours prior to surgery irrespective of the age. Care should be taken to monitor for PE in patients with KTS while leg raising for sterilization. Anesthesiologists should consider potential difficulties in managing the airway and avoid neuraxial anesthesia.

Citation: Lo CY, Chen KB, Chen LK, Chiou CS. Massive pulmonary embolism in Klippel-Trenaunay syndrome after leg raising: A case report. *World J Clin Cases* 2023; 11(17): 4133-4141

URL: <https://www.wjgnet.com/2307-8960/full/v11/i17/4133.htm>

DOI: <https://dx.doi.org/10.12998/wjcc.v11.i17.4133>

INTRODUCTION

Klippel-Trenaunay syndrome (KTS) is a rare congenital disorder and diagnosis of KTS is established when two of the triad features, viz. capillary malformation (port wine stains), hypertrophy of soft tissue or bones and varicose veins, are present. Vascular malformations involve the lower limbs, gastrointestinal tract, and genitourinary tract. The incidence has been estimated at two to five per 100000[1]. KTS predisposes patients to hypercoagulable states, including venous thromboembolism (VTE) and pulmonary embolism (PE). Dislodgement of the clots from pre-existing thrombosed veins upon movements and subsequent migration to pulmonary circulation can lead to massive pulmonary embolism. There have been no reports of severe pulmonary embolisms occurring after leg sterilization, except in cases where a massage or ultrasound examination was being performed.

CASE PRESENTATION

Chief complaints

A 12-year-old girl with KTS (height, 124 cm; weight, 20 kg) was referred to our hospital for recurrent left lower limb lymphangioma since the age of eight years.

History of present illness

She presented with clitoral hypertrophy, hemihypertrophy in the left lower limb along with varicose veins and multiple lipomas, and gastrointestinal bleeding. She was admitted for excision of verrucous hyperkeratosis on the left foot and posterior aspect of the left leg and thigh and excision of a cutaneous hemangioma on the right buttock.

Before induction, the patient was irritably crying in her father's arms at the time. The patient's vital signs were as follows: Heart rate, 111 beats/min; blood pressure, 127/75 mmHg; saturation, 100%; respiratory rate, 18 breaths/min; and body temperature, 36.2°C. After preoxygenation, lidocaine (20 mg), propofol (50 mg), and cisatracurium (4 mg) were administered, and a 5.5 mm cuffed endotracheal tube was inserted. Monitoring was established using standard monitors and an intra-arterial line. Anesthesia was maintained with 45% oxygen and 2% sevoflurane.

After induction, the vital signs were as follows: Saturation, 100%; blood pressure, 97/61 mmHg; heart rate, 113 beats/min; respiratory rate, 17 breaths/min; tidal volume, 200 mL (controlled by ventilator); and body temperature, 36.2°C. Ten minutes after induction, her baseline blood gas test showed pCO₂, 26 mmHg; pO₂, 216.2 mmHg; HCO₃⁻, 19.0 mmol/L; and Hb, 8.4. Sixty minutes after induction, the surgeon elevated the patient's leg for sterilization. Her end-tidal CO₂ dropped from 35 to 7 mmHg within a few minutes. She had hypotension and bradycardia; therefore, atropine and epinephrine were sequentially administered intravenously. The ABG analysis was repeated, and testing revealed acidosis (pH, 7.263; pCO₂, 42.4 mmHg; pO₂, 69.4 mmHg; HCO₃⁻, 19.4 mmol/L; lactate, 1.2 mmol/L; and Ca²⁺, 1.04 mmol/L). Sixty-three minutes after induction, the patient have a cardiac arrest and cardiopulmonary cerebral resuscitation (CPCR) was started. Seventy-eight minutes after induction, a return of spontaneous circulation occurred, and transesophageal echocardiography (TEE) was performed. TEE revealed severe right ventricular distension and little blood flow to the right and left pulmonary arteries. Eighty-six minutes after induction, the patient had a second cardiac arrest and CPCR was

started again. Ninety minutes after induction, a right femoral 5 French-size two-way central line was placed at 13 cm. A cardiac surgeon was consulted who conducted ECMO 106 min after induction. Thirty minutes after ECMO, a normal sinus rhythm returned. CPR was performed for approximately 70 min. The cardiac surgeon started target temperature management (TTM) of 32°C to protect the patient's brain. Due to the incident of severe PE, the surgery was abandoned. Before getting transferred to the pediatric intensive care unit (PICU), her vital signs were as follows: Heart rate, 112 beats/min; blood pressure, 119/100 mmHg; saturation, 100%; respiratory rate, 17 breaths/min; and body temperature, 31.2°C (**Figure 1**).

Initially, in the PICU, her bilateral pupil was dilated approximately 9 mm with no light reflex. She was under sedation with a midazolam pump, as well as a dopamine (10 mcg/kg/min) and heparin pump. Computed tomography (CT) performed three hours after the surgery revealed PEs in the left truncus anterior, right interlobar, bilateral lower lobar, and segmental pulmonary arteries (**Figure 2**). Laboratory data showed elevated D-dimer (7939.76 ng/mL) and decreased fibrinogen (203.3 mg/dL) levels. Six hours later, her body temperature had risen from 31.2°C to 35°C, and her bilateral pupil size was 5 mm with a light reflex. A cardiologist was consulted for thrombolysis six hours after the surgery. Ultrasound-facilitated catheter thrombolysis with the Ekosonic Endovascular System (EKOS) was performed *via* the right femoral vein to the right upper pulmonary artery and left lower pulmonary artery for 48 h from the morning of the first post-incident day (**Figure 3**). On the third post-incident day, a post-procedure CT showed complete resolution of the PEs. The patient was weaned from ECMO on the fifth postoperative day, extubated on the eighth post-incident day, and prophylaxis treatment with enoxaparin was provided. Two months later, the patient was discharged without any neurologic complications (**Figure 4**). The patient had received the surgery 3 months later after the episode.

History of past illness

Four years ago, she had undergone extensive resection of the vascular malformations in the left lower limb, including amputation of the lateral fourth toe on her left foot (**Figure 5**). There was no record of difficulty in intubation.

Personal and family history

There was no specific personal or family history.

Physical examination

Physical examination presented with clitoral hypertrophy, hemihypertrophy in the left lower limb.

Laboratory examinations

Preoperative laboratory data showed anemia (hemoglobin: 9.2 g/dL, mean corpuscular volume: 66.9 fL) no thrombocytopenia or coagulopathy (platelet count, 379000/ μ L; prothrombin time, 11.1 s; international normalized ratio, 1.07; and partial thromboplastin time, 27.9 s), liver and renal function were within normal limit, C-reactive protein: 7.43 mg/L.

Imaging examinations

The preoperative CT revealed angiomatosis extending from the left pelvis to the left foot, pelvic hemangiomas involving the rectum, a splenic lymphangioma, and engorged epidural vessels on the left side of the sacral canal. There is no obvious deep vein thrombosis (DVT) on the preoperative CT.

FINAL DIAGNOSIS

Klippel-Trenaunay syndrome complicated by refractory pulmonary embolism.

TREATMENT

The patient underwent ECMO and received ultrasound-facilitated catheter thrombolysis with the EKOS.

OUTCOME AND FOLLOW-UP

The patient was discharged without any neurologic complications.

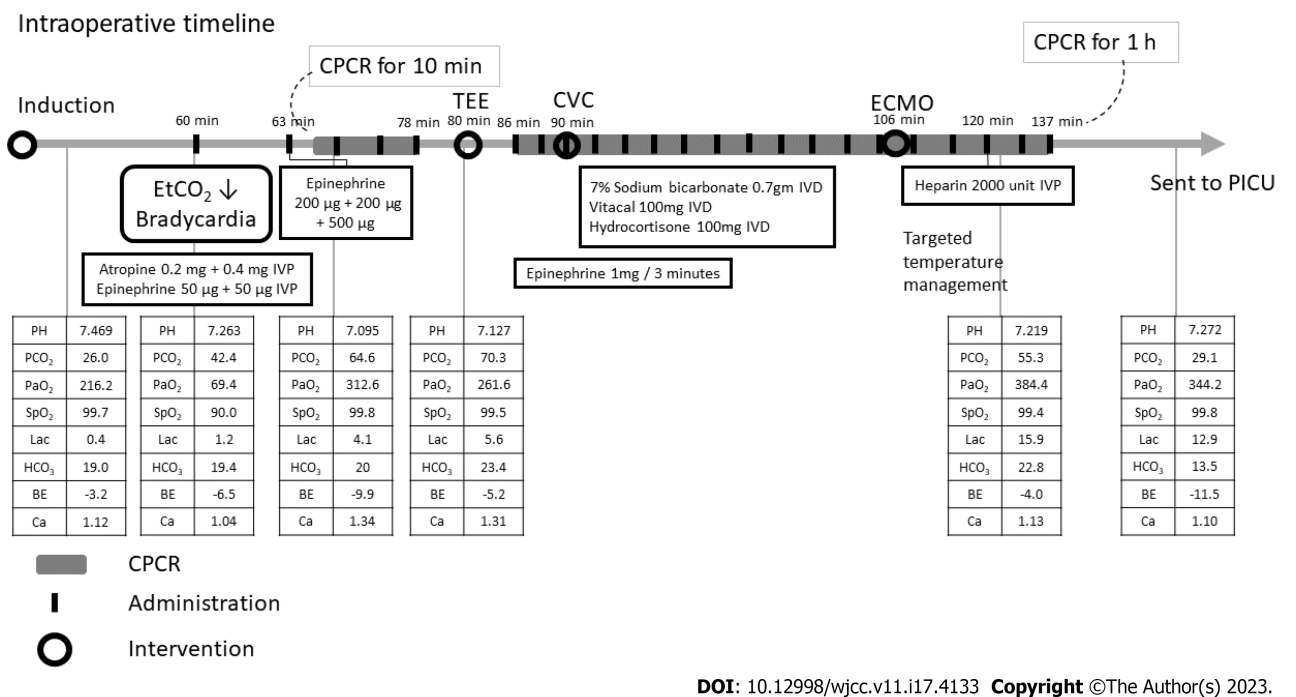


Figure 1 The intraoperative timeline for the patient. CVC: Central intravenous; CTPA: Computed tomography pulmonary angiography; CPR: Cardiopulmonary cerebral resuscitation; ECMO: Extracorporeal membrane oxygenation; TEE: Trans-esophageal echocardiogram; PICU: Pediatric intensive care unit; EtCO₂: End-tidal carbon dioxide.

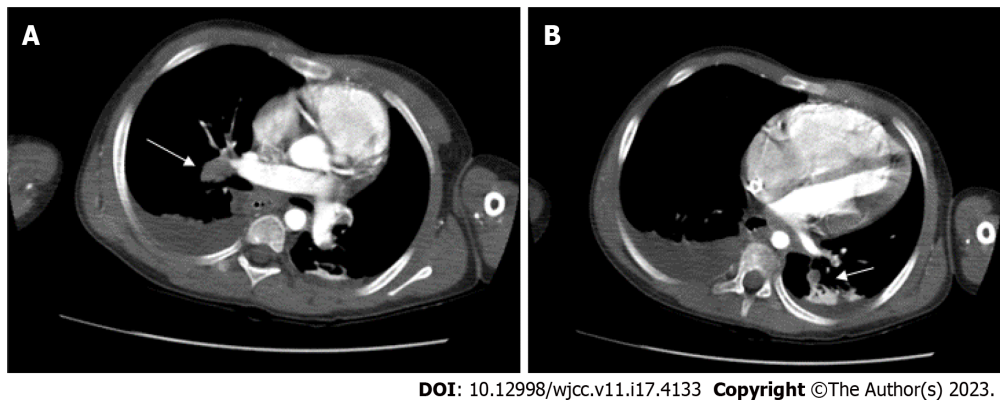


Figure 2 Pulmonary artery embolism. The intraluminal filling defects are noted in A: The right interlobar; B: The left truncus. Bilateral posterior pleural effusions are noted with posterior passive atelectasis of both lungs.

DISCUSSION

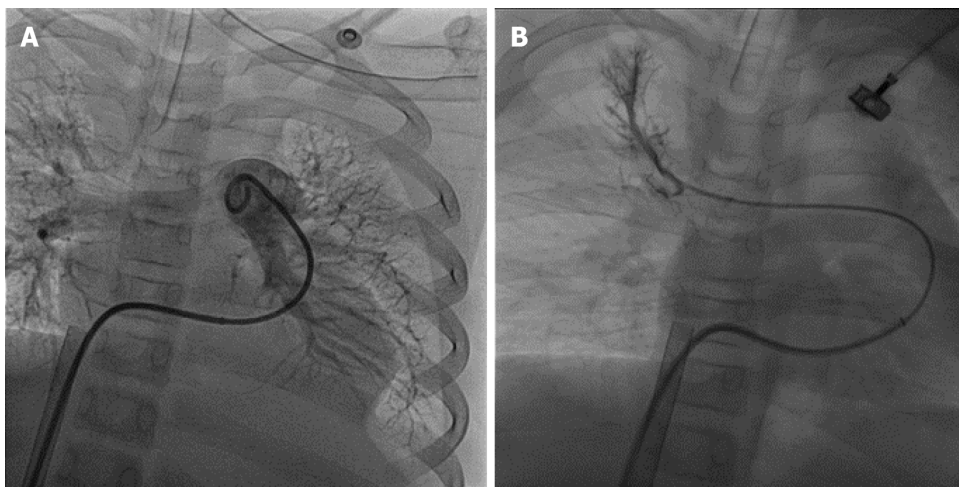
KTS was first described by two French doctors, Klippel and Trenaunay (1900). KTS is a congenital vascular disorder characterized by a triad of main symptoms affecting one or more limbs, namely cutaneous hemangiomas, varicose veins, and bone and soft tissue hypertrophy. Approximately 95% of patients have lower limb malformations[2]. If capillary malformations are sufficiently large, the cutaneous lesions may sequester platelets[3]. In addition, large varicose veins may cause localized intravascular coagulation, and many patients have hematologic evidence of coagulopathy, defined by elevated D-dimer and soluble fibrin complex levels, decreased fibrinogen levels, and elevated prothrombin times, with normal to moderately low platelet counts[4]. Therefore, patients with KTS have a known significantly increased risk for PE[5].

PE while leg raising for sterilization before surgery is rare. The mechanism of PE is that a preexisting deep vein thrombosis is mechanically dislodged by compression or changing positions and travels to the pulmonary artery. Patients can be stratified before surgery based on the clinical prediction scoring system, the Modified Well's criteria (Table 1) recommended by the European Society of Cardiology and the American College of Chest Physicians (ACCP), which standardizes the diagnosis and management of acute PEs. Our patient's score of the Modified Well's criteria is 1.5. However, the Modified Wells'

Table 1 The Clinical prediction scoring system

Modified well's criteria	Points
Clinical symptoms of DVT	3
PE the most likely diagnosis	3
Heart rate > 100/min	1.5
Immobilization or surgery with 4 weeks	1.5
Previous DVT or PE	1.5
Hemoptysis	1
Malignancy	1

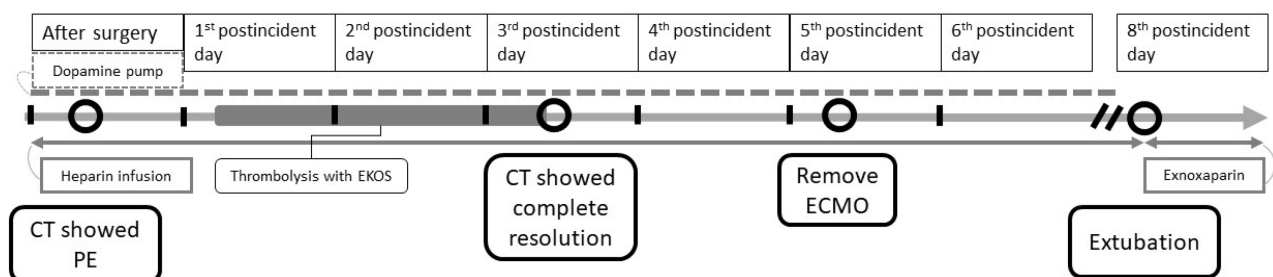
Scoring: < 2: Low probability; 2-6: Intermediate probability; > 6: High probability. DVT: Deep vein thrombosis; PE: Pulmonary embolism.



DOI: 10.12998/wjcc.v11.i17.4133 Copyright ©The Author(s) 2023.

Figure 3 Catheter thrombolysis. Ekosonic Endovascular System-directed thrombolysis was advanced to, A: The left truncus; B: The right upper pulmonary artery.

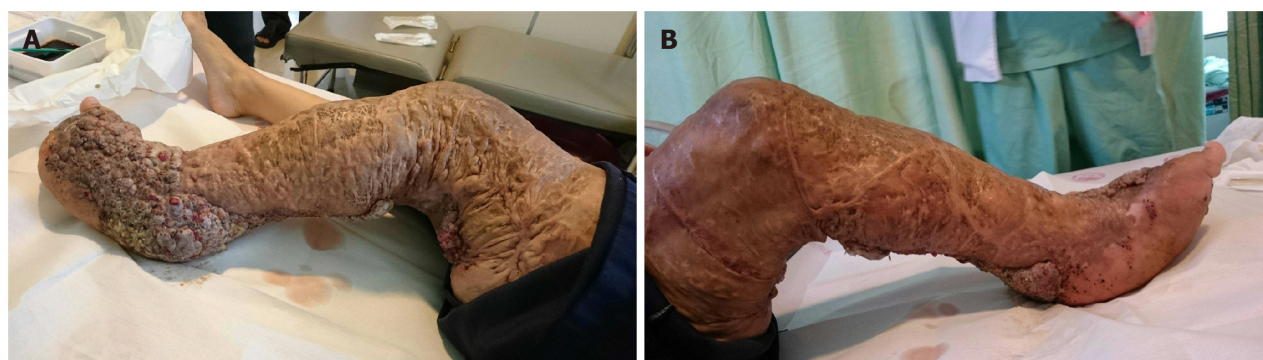
Post-incident timeline



DOI: 10.12998/wjcc.v11.i17.4133 Copyright ©The Author(s) 2023.

Figure 4 The post-incident timeline for the patient. CT: Computed tomography; ECMO: Extracorporeal membrane oxygenation; PE: Pulmonary embolism.

criteria may not be applicable to patients with KTS, as the score may not accurately reflect the patient's high risk for pulmonary embolism[6]. Patients with KTS are at high risk and require a series of examinations, such as ultrasonography, CT, or magnetic resonance imaging. In some case reports[7-9], the thrombi can dislodge from the vein by leg massage, probe compression, and application of sterile elastic exsanguination tourniquets. If thromboembolism is suspected, prevention using anticoagulants and inferior vena cava (IVC) filters might be indicated. However, prophylactic anticoagulants are not prescribed in some Asian regions, and studies have reported a lower incidence of VTE in the Asian population than in Caucasians[10,11]. Although data on pediatric thromboembolism prophylaxis are limited, a systemic review demonstrated that low-molecular-weight heparin is safe and effective in



DOI: 10.12998/wjcc.v11.i17.4133 Copyright ©The Author(s) 2023.

Figure 5 Clinical photograph of the patient showing hypertrophy of the left lower limb. A: The lateral view; B: The medial view.

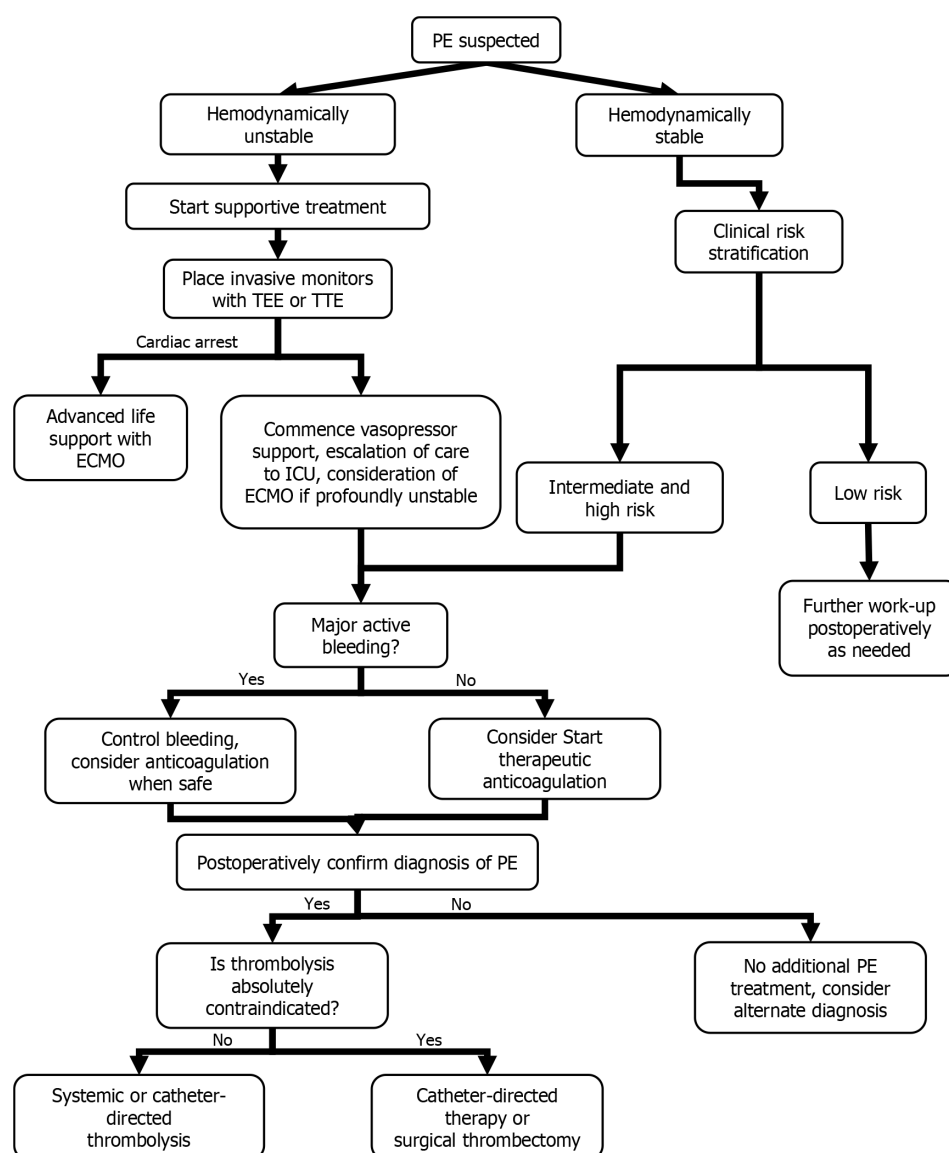
children[12]. The IVC filter is ineffective in KTS because of an anomalous venous connection between the lower extremities and the IVC, effectively bypassing the filter[13]. Considering the risk of lower limb thrombophlebitis associated with venous anomalies, femoral cannulation is better avoided[14].

If PE is suspected intraoperatively, supportive care should be started in unstable patients, including ventilation with 100% O₂, use of positive end-expiratory pressure, resuscitation of the circulating fluid volume, and administration of inotropic drugs as early as possible. In addition, invasive monitors should be placed for diagnosis and management, including arterial and central venous lines, transthoracic echocardiography, and trans-esophageal echocardiogram, which is generally considered the primary diagnostic technique for identifying intraoperative PE because of its high safety, availability, and utility in the operating room and its lack of interference with resuscitation efforts[15]. If vital signs are profoundly unstable, cardiopulmonary cerebral resuscitation should be performed, and a cardiac surgeon should be consulted for cardiopulmonary bypass or venoarterial ECMO. Extracorporeal life support is an effective therapy for unstable patients, offering acceptable outcomes; however, these studies lacked pediatric patients. The American Heart Association guidelines, updated for pediatric advanced life support in 2019[16], recommend ECPR for pediatric patients with cardiac diagnoses and an in-hospital cardiac arrest in settings with existing ECMO protocols, expertise, and equipment. For infants and children who remain comatose after out-of-hospital or in-hospital cardiac arrest, either TTM from 32°C to 34°C followed by TTM of 36°C to 37.5°C or TTM of 36°C to 37.5°C can be used. There were no significant differences in the outcomes between the two TTM groups in the Therapeutic Hypothermia after Pediatric Cardiac Arrest trials[17].

Postoperatively, the patient should be admitted to the intensive care unit, and CT pulmonary angiography should be performed. If there is no major bleeding, anticoagulant treatment should be started as soon as possible, and experts should be consulted for systemic or catheter-directed thrombolysis.

In current standard therapy, if the patient has no major active bleeding, the ACCP recommends immediate initiation of parenteral anticoagulation using low-molecular-weight heparin and fondaparinux, as they are superior to intravenous unfractionated heparin, owing to a lower risk of adverse bleeding events and the absence of need for serial laboratory monitoring. However, there are no guidelines or group consensus for therapies for intraoperative PE. Therefore, the authors organized a reference algorithm for intraoperative PE management (Figure 6).

Despite the lack of established strategies for anesthetic management of KTS patients undergoing surgery, some preoperative evaluations have been recommended. Firstly, anesthesiologists should consider the potential difficulties in managing the airway due to soft tissue hypertrophy in the mouth, hypopharynx, and facial anomalies commonly observed in KTS patients. Secondly, neuraxial anesthesia should be avoided as it is contraindicated due to the possibility of neurovascular malformations in the spinal cord and surrounding structures. It must be noted that central regional blockade can be performed safely in KTS patients, provided that vascular malformations in the central nervous system have been ruled out through computed tomography/magnetic resonance imaging and that there are no cutaneous lesions at the site of needle insertion. Thirdly, the risk of excessive intraoperative blood loss should always be taken into consideration, even in minor surgeries, due to the presence of widespread varicosities and venous malformations. Fourthly, KTS patients have a relatively high risk of developing venous thrombosis and pulmonary thromboembolism. Chronic coagulopathy such as disseminated intravascular coagulation can also occur in these patients. Finally, intracerebral aneurysm is a potential complication in KTS patients, which can rupture during the perioperative period[14,18,19].



DOI: 10.12998/wjcc.v11.i17.4133 Copyright ©The Author(s) 2023.

Figure 6 Algorithm for effective intraoperative decision-making while managing a patient with a pulmonary embolism. ECMO: Extracorporeal membrane oxygenation; TEE: Trans-esophageal echocardiogram; TTE: transthoracic echocardiography; PE: Pulmonary embolism; ICU: Intensive care unit.

CONCLUSION

KTS is a congenital vascular disorder that primarily affects the lower limbs and is characterized by cutaneous hemangiomas, varicose veins, and bone and soft tissue hypertrophy. Patients with KTS are at high risk for PE. All patient having KTS should be evaluated of lower limb circulation and pre-existing DVT pre-operatively. Also, all patients with KTS should receive DVT prophylaxis at least 8 h prior to surgery irrespective of the age. The Modified Wells' criteria may not be applicable to patients with KTS. Care should be taken to monitor for PE in patients with KTS while leg raising for sterilization. Intraoperative PE is lethal, and the management of hemodynamically unstable patients requires efficient CPR and early mechanical support. Anesthesiologists should consider potential difficulties in managing the airway and avoid neuraxial anesthesia. Central regional blockade can be performed safely in KTS patients if vascular malformations in the central nervous system have been ruled out.

FOOTNOTES

Author contributions: Lo CY and Chen KB are the first authors; Lo CY and Chen KB conceptualized and drafted the initial manuscript, and reviewed and revised the manuscript; Chiou CS and Chen LK conceptualized and coordinated, reviewed, and revised the manuscript; All authors approved the final manuscript as submitted and agree to be accountable for all aspects of the work.

Informed consent statement: All study participants, or their legal guardian, provided informed written consent prior to study enrollment.

Conflict-of-interest statement: The authors have no conflicts of interest to disclose.

CARE Checklist (2016) statement: The authors have read the CARE Checklist (2016), and the manuscript was prepared and revised according to the CARE Checklist (2016).

Open-Access: This article is an open-access article that was selected by an in-house editor and fully peer-reviewed by external reviewers. It is distributed in accordance with the Creative Commons Attribution NonCommercial (CC BY-NC 4.0) license, which permits others to distribute, remix, adapt, build upon this work non-commercially, and license their derivative works on different terms, provided the original work is properly cited and the use is non-commercial. See: <https://creativecommons.org/licenses/by-nc/4.0/>

Country/Territory of origin: Taiwan

ORCID number: Chih-Yu Lo 0009-0000-7014-1915; Chiuan-Shiou Chiou 0000-0002-4779-3859.

S-Editor: Liu JH

L-Editor: A

P-Editor: Zhao S

REFERENCES

- 1 Alwalid O, Makamure J, Cheng QG, Wu WJ, Yang C, Samran E, Han P, Liang HM. Radiological Aspect of Klippel-Trénaunay Syndrome: A Case Series With Review of Literature. *Curr Med Sci* 2018; **38**: 925-931 [PMID: 30341531 DOI: 10.1007/s11596-018-1964-4]
- 2 Jacob AG, Driscoll DJ, Shaughnessy WJ, Stanson AW, Clay RP, Gloviczki P. Klippel-Trénaunay syndrome: spectrum and management. *Mayo Clin Proc* 1998; **73**: 28-36 [PMID: 9443675 DOI: 10.1016/s0025-6196(11)63615-x]
- 3 Cha SH, Romeo MA, Neutze JA. Visceral manifestations of Klippel-Trénaunay syndrome. *Radiographics* 2005; **25**: 1694-1697 [PMID: 16284144 DOI: 10.1148/rg.256055042]
- 4 Mazoyer E, Enjolras O, Laurian C, Houdart E, Drouet L. Coagulation abnormalities associated with extensive venous malformations of the limbs: differentiation from Kasabach-Merritt syndrome. *Clin Lab Haematol* 2002; **24**: 243-251 [PMID: 12181029 DOI: 10.1046/j.1365-2257.2002.00447.x]
- 5 Samuel M, Spitz L. Klippel-Trenaunay syndrome: clinical features, complications and management in children. *Br J Surg* 1995; **82**: 757-761 [PMID: 7542989 DOI: 10.1002/bjs.1800820615]
- 6 Ndzengue A, Rafal RB, Balmir S, Rai DB, Jaffe EA. Klippel-trenaunay syndrome: an often overlooked risk factor for venous thromboembolic disease. *Int J Angiol* 2012; **21**: 233-236 [PMID: 24293983 DOI: 10.1055/s-0032-1328969]
- 7 Sutham K, Na-Nan S, Paiboonsithiwong S, Chaksawat P, Tongsong T. Leg massage during pregnancy with unrecognized deep vein thrombosis could be life threatening: a case report. *BMC Pregnancy Childbirth* 2020; **20**: 237 [PMID: 32321459 DOI: 10.1186/s12884-020-02924-w]
- 8 Sufian S, Arnez A, Lakhanpal S. Case of the disappearing heat-induced thrombus causing pulmonary embolism during ultrasound evaluation. *J Vasc Surg* 2012; **55**: 529-531 [PMID: 21958568 DOI: 10.1016/j.jvs.2011.07.070]
- 9 Feldman V, Biadsi A, Slavin O, Kish B, Tauber I, Nyska M, Brin YS. Pulmonary Embolism After Application of a Sterile Elastic Exsanguination Tourniquet. *Orthopedics* 2015; **38**: e1160-e1163 [PMID: 26652340 DOI: 10.3928/01477447-20151123-08]
- 10 Lee CH, Lin LJ, Cheng CL, Kao Yang YH, Chen JY, Tsai LM. Incidence and cumulative recurrence rates of venous thromboembolism in the Taiwanese population. *J Thromb Haemost* 2010; **8**: 1515-1523 [PMID: 20345707 DOI: 10.1111/j.1538-7836.2010.03873.x]
- 11 Liew NC, Alemany GV, Angchaisuksiri P, Bang SM, Choi G, DE Silva DA, Hong JM, Lee L, Li YJ, Rajamoney GN, Suviraj J, Tan TC, Tse E, Teo LT, Visperas J, Wong RS, Lee LH. Asian venous thromboembolism guidelines: updated recommendations for the prevention of venous thromboembolism. *Int Angiol* 2017; **36**: 1-20 [PMID: 27606807 DOI: 10.23736/S0392-9590.16.03765-2]
- 12 Klaassen ILM, Sol JJ, Suijker MH, Fijnvandraat K, van de Wetering MD, Heleen van Ommen C. Are low-molecular-weight heparins safe and effective in children? A systematic review. *Blood Rev* 2019; **33**: 33-42 [PMID: 30041977 DOI: 10.1016/j.blre.2018.06.003]
- 13 Huiras EE, Barnes CJ, Eichenfield LF, Pelech AN, Drolet BA. Pulmonary thromboembolism associated with Klippel-Trenaunay syndrome. *Pediatrics* 2005; **116**: e596-e600 [PMID: 16166386 DOI: 10.1542/peds.2004-1607]
- 14 George SE, Sreevidya A, Asokan A, Mahadevan V. Klippel Trenaunay syndrome and the anaesthesiologist. *Indian J Anaesth* 2014; **58**: 775-777 [PMID: 25624555 DOI: 10.4103/0019-5049.147161]
- 15 Dudaryk R, Benitez Lopez J, Louro J. Diagnosis and Thrombolytic Management of Massive Intraoperative Pulmonary Embolism Guided by Point of Care Transthoracic Echocardiography. *Case Rep Anesthesiol* 2018; **2018**: 8709026 [PMID: 29686907 DOI: 10.1155/2018/8709026]
- 16 Duff JP, Topjian AA, Berg MD, Chan M, Haskell SE, Joyner BL Jr, Lasa JJ, Ley SJ, Raymond TT, Sutton RM, Hazinski MF, Atkins DL. 2019 American Heart Association Focused Update on Pediatric Advanced Life Support: An Update to the American Heart Association Guidelines for Cardiopulmonary Resuscitation and Emergency Cardiovascular Care.

- Circulation* 2019; **140**: e904-e914 [PMID: [31722551](#) DOI: [10.1161/cir.0000000000000731](#)]
- 17 **Moler FW**, Silverstein FS, Holubkov R, Slomine BS, Christensen JR, Nadkarni VM, Meert KL, Browning B, Pemberton VL, Page K, Gildea MR, Scholefield BR, Shankaran S, Hutchison JS, Berger JT, Ofori-Amanfo G, Newth CJ, Topjian A, Bennett KS, Koch JD, Pham N, Chanani NK, Pineda JA, Harrison R, Dalton HJ, Alten J, Schleien CL, Goodman DM, Zimmerman JJ, Bhalala US, Schwarz AJ, Porter MB, Shah S, Fink EL, McQuillen P, Wu T, Skellett S, Thomas NJ, Nowak JE, Baines PB, Pappachan J, Mathur M, Lloyd E, van der Jagt EW, Dobyns EL, Meyer MT, Sanders RC Jr, Clark AE, Dean JM; THAPCA Trial Investigators. Therapeutic Hypothermia after In-Hospital Cardiac Arrest in Children. *N Engl J Med* 2017; **376**: 318-329 [PMID: [28118559](#) DOI: [10.1056/NEJMoa1610493](#)]
- 18 **Lee JH**, Chung HU, Lee MS. An anesthetic management of a patient with Klippel-Trenaunay syndrome. *Korean J Anesthesiol* 2012; **63**: 90-91 [PMID: [22870375](#) DOI: [10.4097/kjae.2012.63.1.90](#)]
- 19 **Barbara DW**, Wilson JL. Anesthesia for surgery related to Klippel-Trenaunay syndrome: a review of 136 anesthetics. *Anesth Analg* 2011; **113**: 98-102 [PMID: [21467557](#) DOI: [10.1213/ANE.0b013e31821a03c2](#)]



Published by **Baishideng Publishing Group Inc**
7041 Koll Center Parkway, Suite 160, Pleasanton, CA 94566, USA

Telephone: +1-925-3991568

E-mail: bpgoffice@wjgnet.com

Help Desk: <https://www.f6publishing.com/helpdesk>

<https://www.wjgnet.com>

