Endarterectomy of the pulmonary artery in patients with myeloproliferative neoplasms

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ABSTRACT

A Pulmonary Thromboendarterectomy (PTE), also known as a Pulmonary Endarterectomy (PEA), is a thoracic surgical procedure that removes clots of blood from the pulmonary arteries, which feed blood to the lungs. Patients with surgically accessible pulmonary artery emboli should have surgery. Recurrent/chronic pulmonary emboli, and hence Chronic Thromboembolic Pulmonary Hypertension (CTEPH), are mainly caused by thrombi. PTE is the only therapy option for CTEPH that has been proven to be effective. Patients with major hemodynamic or breathing issues or impairments may be unable to undergo PTE due to the nature of the technique.

As of 2008, Stuart W. Jamieson of the UCSD Medical Center's cardiothoracic surgery department was widely regarded as a pioneer in the relatively new surgery, having performed more PTEs than the rest of the world combined (over 3000 since 1970 out of a total of 4500 worldwide) with the lowest mortality rate. PTE is exclusively available in the United Kingdom at Royal Papwor Hospital, which is directed by surgeon Mr. David Jenkins. He is o of only four surgeons in the UK qualified to perform pulmona endarterectomy surgery, and they are all based at Royal Papwor Hospital, which is one of the busiest centers in the world for t procedure, with about 190 operations per year and a total caselo of more than 2,000 since 1996.

Myeloproliferative Neoplasms (MPNs) are a type of rare blo malignancies in which the bone marrow produces too many r blood cells, white blood cells, or platelets. Myelo refers to the bo marrow, proliferative to the rapid proliferation of blood cells, a neoplasm to the abnormal and uncontrolled growth of blood cel Overproduction of blood cells is frequently linked to a soma mutation, such as those found in the JAK2, CALR, TET2, a MPL genes. Some MPNs, such as primary myelofibrosis, c accelerate and progress to acute myeloid leukemia in rare situatio

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COMMENTARY

MPNs develop when myeloid lineage precursor cells (blast cells) in the bone marrow get somatic abnormalities that lead them to grow abnormally. The lymphoproliferative disorders, which include acute lymphoblastic leukemia, lymphomas, chronic lymphocytic leukemia, and multiple myeloma, are related diseases in the lymphoid lineage. Genetics is thought to play a key role in the progression of MPNs, particularly in the development of thromboembolic and bleeding problems. Hematologist William Dameshek first introduced the notion of myeloproliferative illness in 1951. The identification of a link between MPNs and the JAK2 gene marker in 2005 and the CALR gene marker in 2013 enhanced MPN classification. In 2008, the World Health Organization classified MPNs as blood malignancies. They were previously termed as Myeloproliferative Disorders (MPD). Mastocytosis was no longer classified as an MPN in 2016.

MPNs are becoming more common, even though they are still considered rare diseases. In some cases, the rate of occurrence has tripled. The rise could be attributed to enhanced diagnostic abilities as a result of the discovery of JAK2 and other gene markers, as well as

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continuing improvement of WHO standards. The incidence and prevalence of MPNs varies widely around the world, with publication bias hypothesised in the cases of essential thrombocythemia and primary myelofibrosis.

Chronic myelogenous leukemia, polycythemia Vera, essential thrombocythemia, and myelofibrosis are all associated with two types of pulmonary hypertension: Chronic Thromboembolic Pulmonary Hypertension (CTEPH) and pre-capillary pulmonary hypertension mimicking pulmonary arterial hypertension. The most effective treatment for CTEPH is Pulmonary Endarterectomy (PEA). However, there are scant data on the outcomes of MPN patients who have had PEA. The goal of this study was to look at the morbidity and mortality rates in CTEPH patients with MPN who had undergone PEA.

All patients scheduled for a PEA between January 2013 and December 2019 was evaluated retrospectively. The protocol was authorized by the Comité d'Ethique pour la Recherche in Anesthésie-Réanimation and registered on Health Data Hub (No. F20210203170207) (IRB 00010254-2020-233). Patients with pulmonary artery obstruction due to sarcoma and emergency procedures were excluded. The diagnosis of MPN was made in accordance with conventional guidelines. In patients with MPN, perioperative anticoagulation was not changed: heparin (anti-Xa activity 0.4-0.6 IU/mL) was started 4 to 6 hours after ICU admission if bleeding was under control.

MPN therapy was reintroduced on the third postoperative day if there were no problems. Infectious complications, the requirement for Extracorporeal Membrane Oxygenation (ECMO) or renal replacement treatment, bronchial artery embolization, mechanical ventilation time, and ICU death were among the postoperative consequences. A clinical suspicion of nosocomial pneumonia was verified by positive bacteriological cultures. Other infections were recorded using the definitions provided by the Centers for Disease Control and Prevention. Hemodynamics was examined two days after PEA before the Swan-Ganz catheter was removed.

The 2 test was used to compare dichotomous variables. If continuous variables were normally distributed, they were represented as mean SD and compared using the Student t test; if not, they were characterized as median (Interquartile Range [IQR]) and compared using the Mann-Whitney U test. The log-rank test was used to compare Kaplan-Meier plots of deaths during ICU stays. Multivariate logistic regression was used to look for risk factors for mortality. Variables linked with P.05 in univariate studies were used in the multivariate model. The median was used to divide the duration of mechanical ventilation. Because age is part of the simplified acute physiology score II, it was not taken into account. The R2 was used to evaluate the model's performance.

PEA was conducted on 675 patients, with 29 (4.3%) of them having MPN (16 polycythemia Vera, 12 essential thrombocythemia, and 1 myelofibrosis). Hydroxycarbamide, pipobroman, anagrelide, and ruxolitinib were used to treat MPN. Patients' characteristics are provided, revealing that MPN patients were older and sicker when admitted to the ICU. PEA had similar early hemodynamic outcomes in participants with and without MPN.

In the ICU, MPN patients had greater infectious problems. There were no differences in the kind of infection across the groups. Nosocomial pneumonia was the most prevalent infectious consequence in both groups (85.4% in patients without MPN vs. 88.9% in patients with MPN, respectively), with identical microbiological findings in both groups (40% Enterobacteriaceae species, 16% Pseudomonas aeruginosa, and 13% Staphylococcus aureus). Greater organ failures were seen in MPN patients, as evidenced by the need for more ECMO and renal replacement treatment.

MPN patients had a five-fold greater ICU death rate. Septic shock, right ventricular failure while on veno-arterial ECMO, hemorrhagic stroke, septic and hemorrhagic shock, and hemorrhagic shock were the leading causes of death. MPN, along with SAPS II score (OR, 1.06; 95% CI, 1.03-1.10; P=.0001) and the need for ECMO (OR, 10.9; 95% CI, 4.29-27.72; P=.0001), was an independent risk factor for ICU mortality (OR, 3.86; 95% CI, 1.15-12.89; P=.028). Renal replacement therapy, viral complications, and mechanical ventilation time were not included as independent variables. The R2 value was 0.36.

According to our findings, MPN is linked to an increased risk of infectious complications and ICU death following PEA. Despite the fact that mortality following PEA was comparable to other studies, our findings show that MPN patients have a higher post-PEA mortality rate. The literature on cardiothoracic surgery in MPN patients is limited, with findings indicating an increased risk of thrombotic and bleeding events but little or no increase in death rates. The poorer outcomes found in our study could be due to a variety of factors. First, MPN patients are more likely to have distal illness, which is the most difficult circumstance to deal with. Second, distinct PH causes, such as CTEPH and concomitant pulmonary microvasculopathy, may coexist in MPN patients.

It's been proposed that the presence and severity of small pulmonary artery dysfunction play a key role in chronic PH after PEA. The increased demand for ECMO in the immediate postoperative period could be due to chronic PH with right ventricular failure or pneumonia-related acute respiratory distress syndrome. Assessing the presence and severity of small pulmonary artery arteriopathy in MPN patients before PEA would be critical. In addition, two-thirds of MPN patients experienced surgical infection problems. When compared to the healthy control population, MPN patients are more susceptible to infection (especially pneumonia) and sepsis, with a higher chance of death as a result. We couldn't locate any data on the rate of surgical infectious problems; however MPN causes chronic inflammation and immune system dysregulation, which can lead to infection. Because the risk of infection is equal in people who receive MPN therapy and those who do not, MPN therapy is likely to be preferred.

There are a few flaws in our research. For starters, this investigation was conducted at a single medical centre. However, because this is a high-volume hospital in charge of PEA on a national level, vast numbers of patients treated in a comparable way by interdisciplinary expert teams can be studied. Second, due to the disease's rarity, the number of cases is still modest. As a result, a prospective multicenter worldwide study could yield more reliable results. Third, no thrombotic or hemorrhagic consequences were recorded. The diagnosis of pulmonary artery rethrombosis, in particular, was not routinely reported.

In comparison to other CTEPH groups, PEA in CTEPH patients with MPN is associated with greater mortality and a higher rate of infectious comorbidities. As a result, such patients require a thorough preoperative multidisciplinary evaluation in a high-volume expert facility.