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ORIGINAL ARTICLE

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Impact of the establishment of a multidisciplinary national chronic thromboembolic pulmonary hypertension board on a monocentric surgical endarterectomy program

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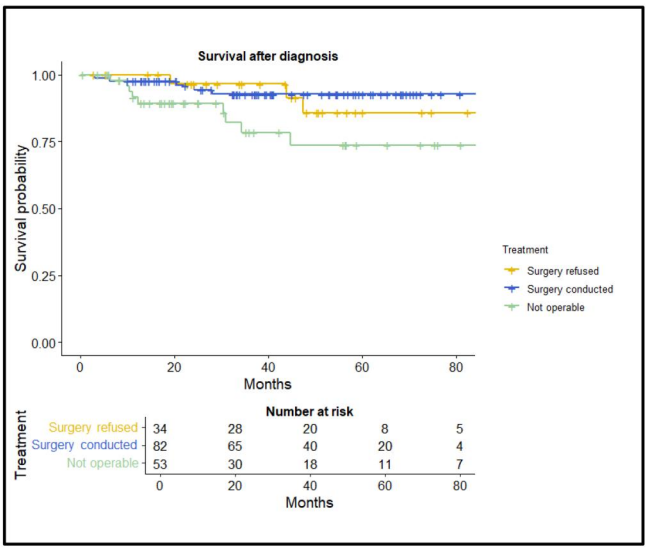
Summary

CTEPH patients were reviewed by our CTEPH Board. Survival rates significantly differ between those undergoing PEA, operable patients refusing PEA, and non-operable patients. The PEA rate rose from 14% to 41% after the CTEPH Board's establishment. The Swiss national CTEPH Board was crucial in increasing curative surgery rates and improving patient survival.



Legend: Chronic Thromboembolic Pulmonary Hypertension (CTEPH), Pulmonary Endarterectomy (PEA)

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Abstract

OBJECTIVES: Chronic thromboembolic pulmonary hypertension is a rare disease, characterized by delays in diagnosis and curative surgical treatment. After establishing a surgical pulmonary endarterectomy centre in Switzerland and due to a historically low resection rate of 14%, a national multidisciplinary evaluation board was established in January 2018. Herein, we summarize the impact of the board on our programme.

METHODS: Patients discussed in the national chronic thromboembolic pulmonary hypertension board from January 2018 to December 2023 were included. Clinical characteristics, treatment allocation and survival were compared between patients undergoing surgery, patients refusing surgery and non-operable patients. Fisher's exact test or three-way ANOVA and Kaplan–Meier analyses were used.

RESULTS: 188 patients were discussed at our national chronic thromboembolic pulmonary hypertension board; 131 (70%) presented with operable disease, 77 (41%) were referred for pulmonary endarterectomy and 34 (18%) of operable patients declined surgery. There is a significant difference in survival between these groups ($P = 0.048$). One- and 2-year survival in the subgroup undergoing pulmonary endarterectomy was 97% and 79%, respectively, while 1- and 2-year survival in the subgroup refusing pulmonary endarterectomy was 91% and 76%, respectively. The pulmonary endarterectomy rate has increased from a historical low of 14–41% since establishing the board.

CONCLUSIONS: Establishing an interdisciplinary board is essential to address diagnostic and management challenges in chronic thromboembolic pulmonary hypertension patients. The Swiss national chronic thromboembolic pulmonary hypertension board played an important role in substantially increasing the rate of curative surgery.

Keywords: chronic thromboembolic pulmonary hypertension • pulmonary endarterectomy • multidisciplinary board

ABBREVIATIONS

BPA	Balloon pulmonary angioplasty
CTEPH	Chronic thromboembolic pulmonary hypertension
ERS	European Respiratory Society
ESC	European Society of Cardiology
IQR	Interquartile range
NYHA	New York Heart Association
PEA	Pulmonary endarterectomy
PH	Pulmonary arterial hypertension
SSPH	Swiss Society of Pulmonary Hypertension

INTRODUCTION

Chronic thromboembolic pulmonary hypertension (CTEPH)—a rare precapillary form of pulmonary hypertension, classified as group 4 of pulmonary hypertension (PH)—is a potentially fatal disease often associated with preceding acute pulmonary embolism and defined by symptomatic PH with persistent perfusion defects despite 3 months of adequate anticoagulation [1, 2]. CTEPH is underdiagnosed and undertreated. Recent international data show an annual incidence of 3–5 cases per 100 000 individuals [3]. Diagnostic delay is a significant issue, with a median time of 14 months from symptom onset to diagnosis [4–7]. This delay is associated with worse haemodynamic profiles and shorter survival rates [4, 8, 9]. Untreated mean pulmonary arterial pressure above 30 mmHg is linked to a 90% 3-year mortality [10, 11]. Pulmonary endarterectomy (PEA) is the gold standard treatment for CTEPH with surgically accessible lesions [2]. It is associated with significantly improved medium- to long-term survival compared with conservative treatment [8]. Balloon pulmonary angioplasty (BPA) is a therapeutic option for inoperable patients or those with residual PH post-PEA. Combining medical therapy with interventional procedures offers a comprehensive approach to managing CTEPH, improving survival and quality of life [2, 8, 12, 13].

According to the international registry and European guidelines, centres performing >50 PEAs per year had the best in-hospital and 1-year mortality rates [2, 14]. Given the country's small population, these PEA rates are not realistic in Switzerland. Until 2015, there was no PEA programme in Switzerland, and therefore from 2000 to 2012, only 14% and from 2011 to 2015, 28% of patients received PEA [15, 16], and patients had to travel to other countries for surgery (Germany, France). After establishing a PEA centre in Zurich, Switzerland, the need for a national multidisciplinary evaluation Board was recognized. Here, we discuss the effect of the establishment of a multidisciplinary national CTEPH Board in Switzerland supported by the Swiss Society of Pulmonary Hypertension (SSPH) on the outcome of Swiss CTEPH patients.

MATERIALS AND METHODS

Ethical statement

The local ethics committee reviewed this project for clarification of responsibility BASEC-ID (Req-2024-01486). This project is classified as quality assurance and, therefore, does not fall within the scope of the Human Research Act and does not require the approval of the competent ethics committee before its implementation.

Patients and methods

The national Swiss CTEPH Board was inaugurated in January 2018, supported by the SSPH. The board comprises a multidisciplinary collaborative team of pulmonologists, cardiologists, radiologists and thoracic surgeons. The Swiss National CTEPH Board includes leading medical centres from across Switzerland. The board meets monthly to discuss patient cases, treatment protocols and research advancements in CTEPH.

The study design is a retrospective cohort study. Patients included were all patients with CTEPH discussed at the CTEPH Board between January 2018 and December 2023. Patients without confirmation of a CTEPH diagnosis were excluded.

We retrospectively divided the patients into three groups: operable patients undergoing PEA, operable patients refusing PEA and non-operable patients. Patients with a follow-up status no longer than 6 months prior to this analysis were included. Extensive efforts were undertaken to minimize the loss of follow-up and missing data including gathering data from the most recent follow-up from referring hospitals, primary care physicians or patients. Follow-up rates were estimated using the simplified person-time method by Xue *et al.* [17]. Patient data were extracted from the hospitals electronic patient system. The data were managed using REDCap electronic data capture tools hosted at the University Hospital of Zurich.

Statistical analysis

Tables and charts were created using Microsoft Excel 2016. The Swiss-map was hand-drawn. The Kaplan–Meier survival curve was created using RStudio (2020; RStudio: Integrated Development for R). The Sankey plot was created using SankeyMATIC open-source tool (2024). The resection rate diagram and logistic regression were done using MatLab MathWorks Inc. (2022). For the descriptive data analysis and presentation, we used total number, percentage and median with Interquartile range (IQR). For categorical data, the *P*-values were calculated with Fisher's exact test, for continuous data with three-way ANOVA and survival with log-rank test. Pairwise *P*-values are reported for survival comparisons, using hazard ratios estimated from a Cox proportional hazards model, which accounts for the log-linear relationship of risks. The significance level was defined at $P = 0.05$. A multinomial logistic regression was used to analyse characteristics associated with undergoing PEA. Missing data and values were reported as such. The outcomes of interest were survival and resection rate compared to the historical cohort.

RESULTS

In total, 188 CTEPH patients were discussed at the CTEPH Board. The patients (Table 1) median age was 68 years (IQR 55–75 years);

Table 1: Baseline characteristics and survival outcomes of patients with CTEPH discussed at the Swiss national CTEPH Board from January 2018 to December 2023

	All CTEPH patients	Non-operable	PEA refused	PEA conducted
Number of cases	188	57	34	77
Age in years				
Median [IQR]	68 [55 75]	72 [61.5 78.25]	74 [59.75 77]	62 [51 70]
Gender				
Male	107 (56.91%)	26 (45.61%)	16 (47.06%)	50 (64.94%)
Female	81 (43.09%)	31 (54.39%)	18 (52.94%)	27 (35.06%)
NYHA				
I	10 (5.32%)	4 (7.02%)	3 (8.82%)	2 (2.60%)
II	72 (38.30%)	23 (40.35%)	14 (41.18%)	24 (31.17%)
III	96 (51.06%)	25 (43.86%)	16 (47.06%)	47 (61.04%)
IV	8 (4.26 %)	4 (7.02%)	–	4 (5.19%)
Unknown	2 (1.06%)	1 (1.75%)	1 (2.94%)	–
BMI				
Median [IQR]	26.29 [23.67 29.77]	25.67 [23.08 29.84]	27.14 [23.79 30.01]	26.64 [23.51 30.09]
Patient status, *n* (%)				
Alive	170 (90.42%)	52 (91.22%)	30 (88.2%)	73 (94.81%)
Dead	18 (9.56%)	5 (8.78%)	4 (11.8%)	4 (5.19%)
1-Year survival after diagnosis, *n* (%)				
Alive	152 (80.85%)	44 (77.19%)	31 (91.18%)	74 (97.37%)
Dead	6 (3.19%)	4 (7.02%)	0 (0%)	2 (2.63%)
Not reached	30 (15.96%)	9 (15.79%)	3 (8.82%)	0 (0%)
2-Year survival after diagnosis, *n* (%)				
Alive	120 (63.83%)	31 (54.39%)	26 (76.47%)	60 (78.95%)
Dead	9 (4.79%)	5 (8.77%)	1 (2.94%)	2 (2.63%)
Not reached	59 (31.38%)	21 (36.84%)	7 (20.59%)	14 (18.42%)
Survival after PEA [months]				
Median [IQR]	Not applicable	Not applicable	Not applicable	36.20 [16.07 53.23]
30-Day mortality, *n* (%)	Not applicable	Not applicable	Not applicable	
Alive				75 (97.40%)
Dead				2 (2.60%)
90-Day mortality, *n* (%)	Not applicable	Not applicable	Not applicable	
Alive				75 (97.40%)
Dead				2 (2.60%)
Hospitalization duration [days]				
Median [IQR]	Not applicable	Not applicable	Not applicable	13 [11 19]
ICU duration [days]				
Median [IQR]	Not applicable	Not applicable	Not applicable	4 [3 7]
In-hospital mortality	Not applicable	Not applicable	Not applicable	2 of 77 (2.60%)

One hundred thirty-one (69.68%) patients were technically operable, 77 (40.9%) underwent PEA in this period and 20 (10.6%) PEAs are still pending at the time of December 2024. The age differences between the groups were significant between patients refusing PEA and undergoing PEA ($P = 0.03$) and between patients undergoing PEA and non-operable ($P = 0.0018$). The difference between genders was significant when comparing non-operable patients and patients undergoing PEA ($P = 0.033$). For the 1- and 2-year survival, 'not reached' refers to the event discussed not having been 1 or 2 years prior to this analysis. ICU: intensive care unit; IQR: interquartile range; NYHA: New York Heart Association.

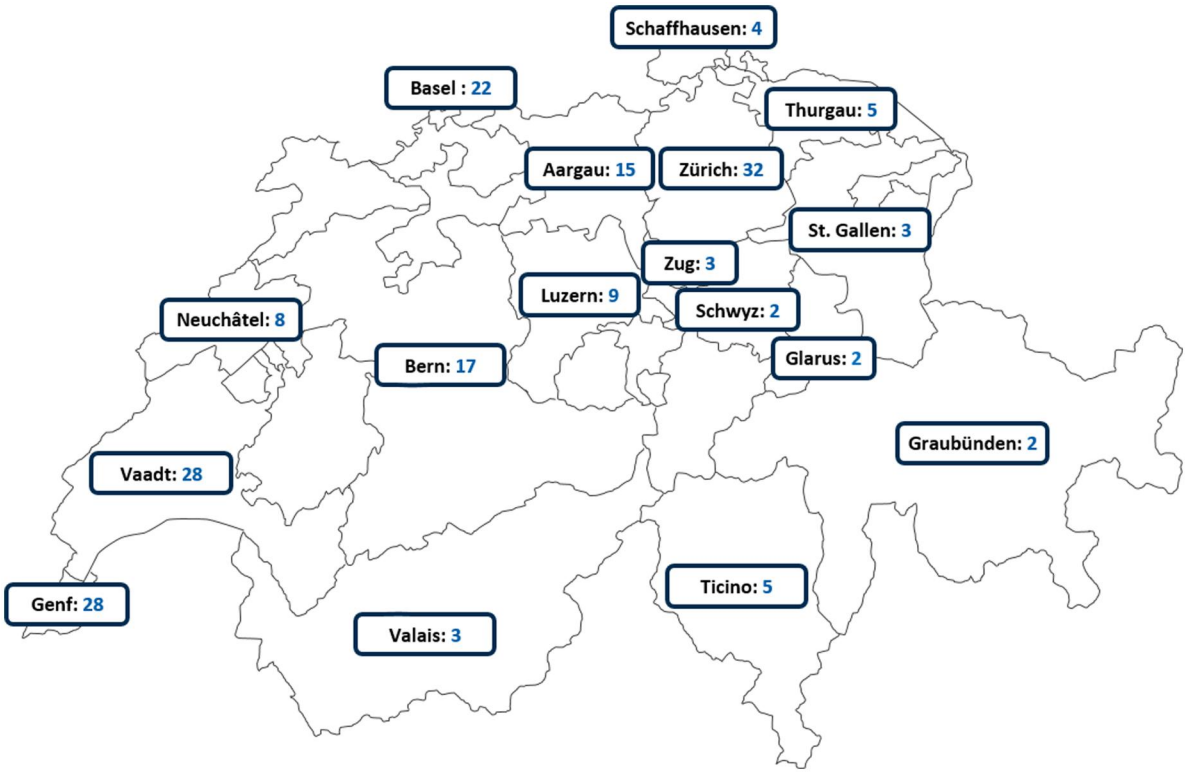

Figure 1: Geographical map of Switzerland indicating cantons, from which patients with CTEPH were referred. Number of referrals for the corresponding canton is indicated

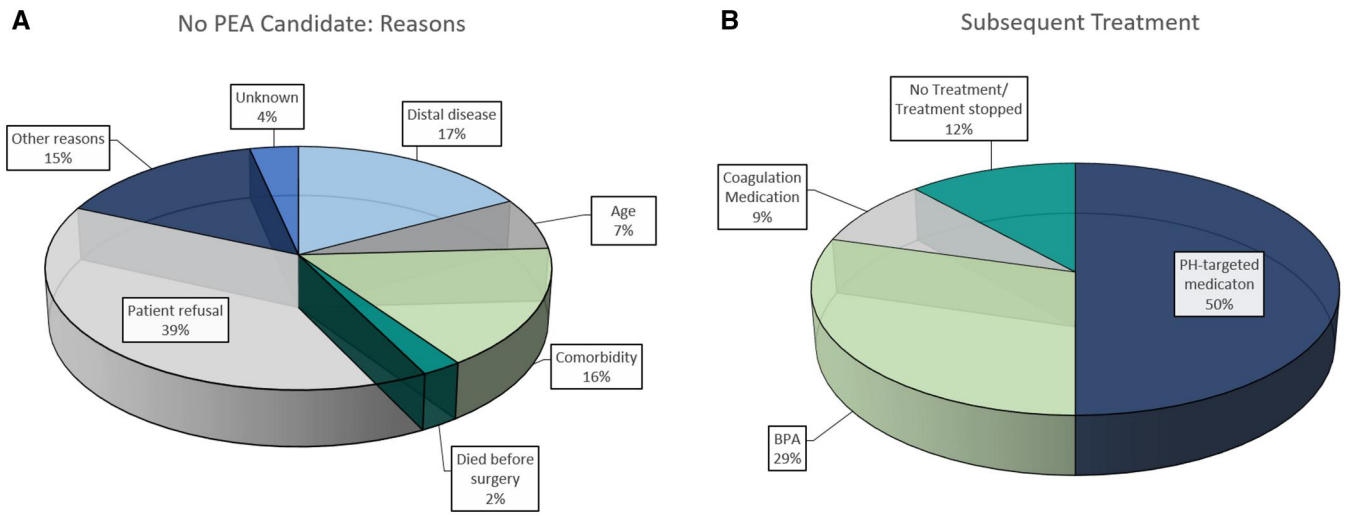

Figure 2: (**A**) Pie-chart illustrating the reasons why patients with CTEPH were classified as non-surgical candidates (proportions represented in percentages). (**B**) Pie-chart showing different alternative types of treatments for patients with operable CTEPH who refused PEA (proportions represented in percentages)

57% of patients were male ($n = 107$). Patients discussed at the CTEPH Board were referred from all over Switzerland (Fig. 1). One hundred thirty-one (70%) patients presented with operable disease, and 77 (40%) received PEA. Altogether, 85 (45%) were classified as no PEA candidates (Fig. 2). Yet, 53 (28%) patients were truly inoperable and 4 (2%) were unknown. Of the 34 patients who refused surgery, 88% ($n = 30$) were still alive at the time of analysis. Their median age when discussed at the Board was 74 years (IQR 59.75–77 years). In this subgroup, 38% had New York Heart Association (NYHA) functional class II and 47% functional class III (Fig. 3). Their subsequent treatments are shown in Fig. 2. In the subgroup of 77 patients who underwent PEA, 73 (95%) were still alive at the time of analysis. Their median age at presentation at the Board was 62 years (IQR 51–70 years). In this subgroup, 31% had NYHA functional class II and 61% had functional class III (Fig. 3). Thirty- and 90-day mortality was $n = 2$ (2.6%).

Using a multinomial logistic regression, we observed that male patients ($P = 0.03$), younger patients ($P = 0.025$) and patients with higher NYHA classification ($P = 0.015$) were more

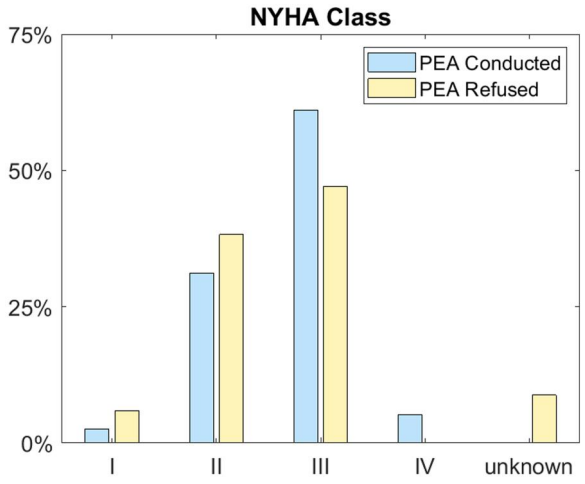

Figure 3: Bar-chart showing NYHA classification for patients with CTEPH who received PEA and who refused the surgery

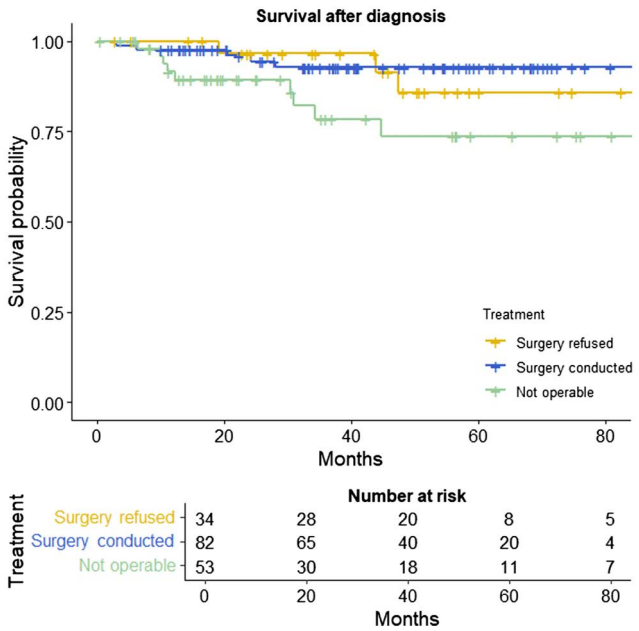

Figure 4: Kaplan–Meier survival curve and numbers at risk comparing survival of PEA candidates who refused the operation, PEA candidates who underwent PEA and patients who were non-operable after diagnosis. Using log-rank test, the differences in OS between these subgroups ($P = 0.048$) was significant. Using Cox proportional hazards model, the difference between surgery conducted and non-operable was significant ($P = 0.02$), the difference between surgery conducted and refused ($P = 0.5$) and between refused and non-operable ($P = 0.3$) not significant. The timeline is presented in months, and survival curves were truncated when less than 10% of the initial sample size remained at risk

likely to undergo PEA. Figure 4 shows Kaplan–Meier survival curves comparing the overall survival of operable patients who underwent PEA with those who refused PEA and non-operable patients. The differences in overall survival between these subgroups were statistically significant ($P = 0.048$), in particular a significant difference between non-operable and patients who underwent PEA ($P = 0.02$). The 1- and 2-year survival in the subgroup undergoing PEA was 97% and 79%, respectively, and, in the subgroup, refusing PEA, 91% and 76%, respectively (Table 1). The median follow-up duration for the entire study cohort was 37.37 months (IQR 19.03–58.63). The follow-up rate was 100% (simplified person-time method).

Multimodal treatment strategy in CTEPH patients is depicted in Fig. 5. Eight (4%) patients received BPA post-PEA, 15 (8%) patients received PH-targeted medication after PEA, 5 (2.6%) after BPA and 17 (9%) patients received PH-targeted medical therapy only.

DISCUSSION

We analysed the demographic and clinical characteristics of 188 patients with a diagnosis of CTEPH discussed at the Swiss National CTEPH Board since its inauguration. The majority (70%) of patients presented with operable disease, of whom 41% underwent PEA. The main reason for patients to be classified as non-PEA candidates was patient refusal ($n = 34$, 18%). Compared to historical data from the Swiss PH registry, we observe a substantial increase in PEAs performed since both the establishment of the CTEPH Board in 2018 and the surgical CTEPH centre in Switzerland in 2015. From 1998 to 2012, 14% and 2% of the reported 249 CTEPH patients underwent PEA or lung transplantation, respectively; 87% were treated with PH-targeted therapy. These patients primarily needed to travel outside the country to have access to curative surgery [16]. In Swiss CTEPH patients, the proportion of patients undergoing PEA increased from 15% in 2001–2005 to 40% in 2016–2019. However, until 2019, only 25% of patients in the Swiss registry have been surgically treated for CTEPH [15]. Therefore, the resection rate increased from 14% to 41% (Fig. 6). An international CTEPH registry reported a resection rate of 58% [18]. In patients undergoing PEA, we observed an in-hospital mortality, 30- and 90-day mortality of 2.6%. Our data are comparable to survival outcomes of high-volume centres, which report <5% in-hospital mortality [14], 4% 30-day mortality [19], 3.9% 90-day mortality [20] and 12.3–1.9% 30-day mortality over the course of 23 years' experience [21].

Patients who were younger at board presentation seemed more likely to undergo PEA. The differences in clinical characteristics between PEA patients and those who were operable but declined surgery suggest that older patients tend to be more hesitant to undergo this complex procedure, whereas younger patients may perceive that long-term benefits of surgery outweigh associated risks. Similar to our observations, in a worldwide CTEPH registry, the median age for patients with CTEPH was 63 years (IQR 51.0–73.0) [18]. As we observed, in various previous reviews, the patient cohorts that undergo PEA also tend to be younger than the general CTEPH cohort [18–21].

Our cohort has a modestly higher prevalence of the male sex, whereas some centres report female dominance in PEA patients [19]. Interestingly, in the worldwide prospective CTEPH registry, sex is evenly balanced, except for Japan (75% female) [18]. We also report a difference between the sexes in the PEA subgroup, where women appear to be less likely to receive PEA. This has also been reported in previous studies, including a recently published European CTEPH registry where fewer women chose to undergo PEA [22–25].

Additionally, higher preoperative NYHA functional class increased the likelihood of undergoing PEA. This suggests that patients opting for PEA may already have more symptoms, a higher disease burden and are more willing to undergo a complex surgery to improve their quality of life. Hence, patients refusing surgery had less symptoms and were therefore less

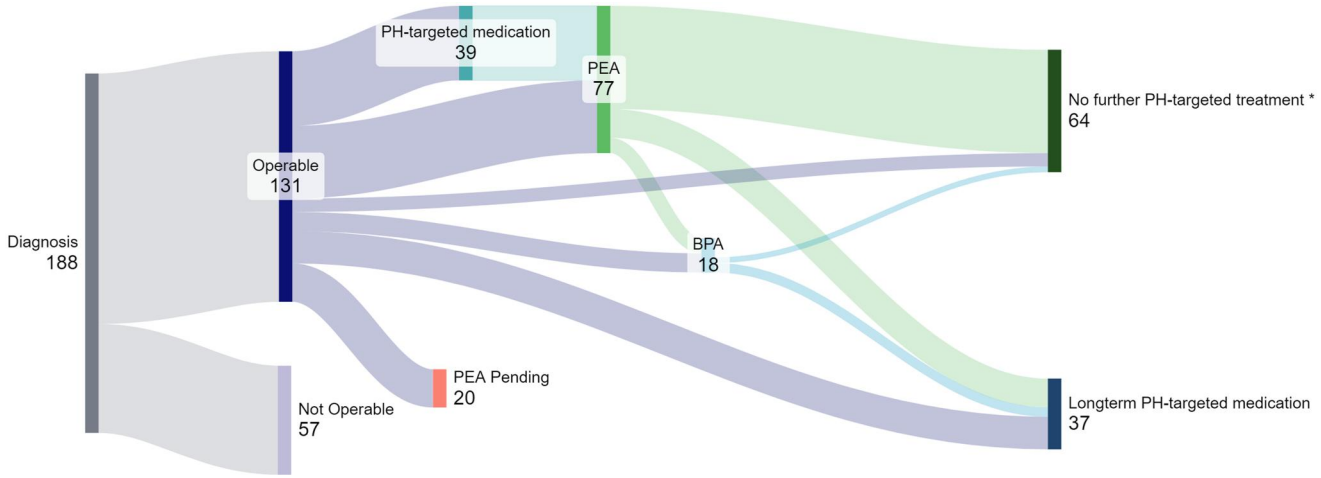

Figure 5: Sankey plot displaying the multimodal treatment of patients with CTEPH discussed at the Swiss national CTEPH Board. One hundred thirty-one (69.6%) patients were operable, 57 (30.3%) patients were non-operable. Thirty-nine (20.74%) patients received PH-targeted medication prior to PEA. Seventy-seven (40.95%) patients underwent PEA. Eighteen (9.57%) operable patients received BPA, including 8 (4.25%) that received a BPA post-PEA. Thirty-seven (19.68%) operable patients received long-term PH-targeted medication. Fifteen (7.97%) patients received PH-targeted medication post-PEA, 5 (2.6%) post-BPA and 17 (9.04%) patients received PH-targeted medication only. Sixty-four (34.04%) operable patients had no further PH-targeted specific treatment*, including 54 (28.72%) patients post-PEA, 3 (1.59%) patients post-BPA and 7 (3.6%) who did not receive either intervention. At the time of this analysis, 20 (10.63%) PEAs are pending

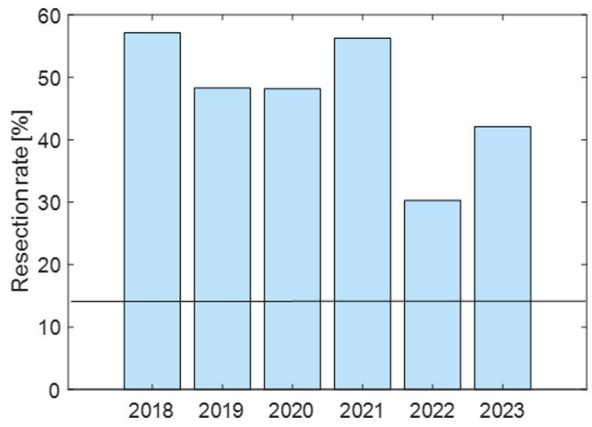

Figure 6: Histogram portraying resection rates for each year since the establishment of the board. 2018 (57.1%), 2019 (48.3%), 2020 (48.2%), 2021 (56.3%), 2022 (30.3%) and 2023 (42.1%). The overall resection rate is 41%. We included a line at the historic resection rate of 14%

motivated to undergo PEA. This is in line with published research, which shows that while all patients present with functional limitations, those who eventually underwent PEA tend to have an even worse preoperative functional class than non-operated patients [19, 20].

The current trend for multimodal treatment is conveyed in this cohort; 21% of patients received PH-targeted medication prior to PEA. After PEA, 4% of patients received a BPA. In total, 15 (8%) patients of the whole cohort received long-term PH-medication after PEA, and 5 (2.6%) patients after BPA. Liberal prescription practices, where we prescribe PH-targeted medication for pulmonary vascular resistance >8 WU and mean pulmonary arterial pressure >25 mmHg, could explain the high rates of multimodal treatment. We also report few patients after PEA, which will proceed to BPA and/or PH-targeted medication treatment, when indicated according to the European Society of Cardiology (ESC)/European Respiratory Society (ERS) guidelines [2]. Treatment with BPA is used for inoperable patients or patients refusing surgery, as well as patients having residual PH after PEA. The multimodality treatment seen in this cohort is in alignment with ECS/ERS guidelines and with the worldwide prospective CTEPH registry, where 58.7% of patients had PEA, 17.3% had BPA and 14.7% had PH-targeted therapy only [2, 18]. Further, an overlap of PH-targeted medication with PEA and BPA candidates was previously reported, with up to one-third of patients receiving targeted PH medication in addition to PEA [18, 19]. Therefore, our cohort portrays the reality of multimodality in clinical practice and highlights the importance of a multidisciplinary teamwork for these complex patients with the need for a tailored therapeutic approach.

The significantly higher rate of 1- and 2-year survival rates in the subgroup of patients undergoing PEA compared to the subgroup refusing PEA is coherent with international data, showing a 92.4% 1-year survival in a UK specialist centre [21] and also with the international registry reporting 1- and 2-year survivals of 93% and 91% in operated patients, and only 88% and 79% in non-operated patients [8]. This highlights an expected greater postoperative risk in PEA patients but with excellent early and long-term results, as it has also been described by de Perrot *et al.* [19]. Additionally, the reported survival in this analysis aligns with the trajectory of improvements in survival in the Swiss PH registry, which reported an improvement in 3-year survival from 86% to 93% in the period from 2001 to 2019 [15].

The establishment of the CTEPH Board also aimed to raise awareness of CTEPH. Other strategies to raise awareness were newspaper articles, presentations at conferences, articles in national journals and television segments. The referral pattern, including all parts of Switzerland, hints at an improved awareness of general practitioners for CTEPH.

Limitations

The study is based on patients from a single country, limiting its generalizability. This analysis does not focus on BPA as standalone primary treatment. A detailed analysis of BPA would be interesting for future research. Future studies are warranted to

analyse the diagnostic delay of patients with CTEPH. Further efforts need to be undertaken to better understand patient refusal, to investigate measures to improve a timely diagnosis and treatment allocation.

CONCLUSION

In summary, the study provides a comprehensive overview of the demographics, functional status, regional distribution and outcome of patients with CTEPH referred to the national interdisciplinary CTEPH Board in Switzerland. Increased awareness has led to a substantial increase in surgical procedures performed. The high proportion of patients in NYHA class II and III suggests that most patients are diagnosed when they are already experiencing significant symptoms, emphasizing the ongoing need for early detection and intervention. The sex-related differences observed highlight a further need for care in understanding why more female patients refuse the curative treatment. Favourable outcomes can be achieved in smaller volume centres with globally comparable mortality and survival data for patients with CTEPH discussed by a multidisciplinary team. The Swiss national CTEPH Board has played an important role in accurate and timely diagnosis and significantly increasing access to curative surgery for Swiss CTEPH patients.

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DATA AVAILABILITY

The data underlying this article will be shared on reasonable request to the corresponding author.

Author contributions

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