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Clinical Implications of Hemoptysis in Patients with Pulmonary Arterial Hypertension

Jose Cantu, MD¹, Degang Wang, PhD², and Zeenat Safdar, MD, FCCP¹

¹Division of Pulmonary-Critical Care Medicine, Baylor College of Medicine, Houston, Texas

²Baylor College of Medicine, Houston VA health Services Research and Development Center of Excellence, Michael E. DeBakey VA Medical Center, Texas, United States

Abstract

Introduction—Pulmonary arterial hypertension (PAH) is a disabling disease that may result in haemoptysis. Patients with congenital heart disease associated PAH (CHD-APAH) may have a survival advantage when compared with patients with other types of PAH presenting with haemoptysis. The effects of aetiology and sub-sequent management choice of haemoptysis in PAH patients is not well-defined.

Methods—We conducted outcome analysis in CHD-APAH vs. all other subtypes of PAH patients presenting with haemoptysis to The Methodist Hospital. Twenty-one patients were identified, thirteen patients in the CHD-APAH group and eight patients in the non-CHD group. We evaluated outcomes related to treatment (bronchial artery embolization vs. conservative management), hospital length of stay, mortality rates and survival in this cohort.

Results—The CHD-APAH and non-CHD groups had similar baseline demographic, haemodynamic and laboratory values except BMI was higher in the non-CHD group and haematocrit was higher in the CHD-APAH group. Twenty-eight-day mortality (0% vs. 31%) and 1-year mortality (0% vs. 54%) was lower in the CHD-APAH patients as compared with non-CHD group. A statistically significant difference was found in the survival rate in favour of CHD-APAH group for the total follow-up period ($p = 0.02$). Although not statistically significant, patients treated with BAE had shorter length of stay ($4.0 \text{ days} \pm 4.0$ vs. $13.7 \text{ days} \pm 22.5$; $p = 0.26$). There was recurrent haemoptysis in 43% of patients treated with BAE.

Conclusion—Haemoptysis in PAH patients is a serious event with a high mortality rate. CHD-APAH seems to confer a survival advantage, independent of therapy utilised. Termination of haemoptysis with BAE is rapid with relatively few complications except for frequent re-bleeding episodes. Further studies are needed to determine the risk factors that may predispose PAH patients to excessive mortality from haemoptysis and to identify an optimal therapeutic modality.

Keywords

brochial artery embolization; right heart failure; echocardiogram

Introduction

Pulmonary arterial hypertension (PAH) is a serious disease that produces progressive obstruction and obliteration of the pulmonary vascular bed leading to death. A variety of systemic consequences of long standing PAH have been identified including hemoptysis.¹ The incidence of hemoptysis in patients with PAH is unknown and is likely underreported in the literature.² Previously published retrospective case reports highlight hemoptysis in PAH patients as a seminal event with excessive mortality.^{3, 4}

Mortality related to hemoptysis is likely mitigated by a multitude of factors. There is some evidence that patients with congenital heart disease related PAH (CHD-APAH) may fare better than patients with other types of PAH.⁵ The therapeutic intervention may also influence subsequent survival. Several strategies have been employed in the past including supportive care, surgical resection and lung transplant. Currently, the most commonly used strategy is bronchial artery embolization (BAE). This technique involves injecting particulate matter into angiographically identified bronchial arteries that are abnormal. Flow ceases through these abnormal vessels in attempt to hinder further bleeding.⁶ However, recurrent bleeding is reported to be common with BAE, although the procedure is well tolerated.^{4,7-9}

In this study, we present data from PAH patients presenting with hemoptysis that was analyzed for trends in hospital length of stay and mortality with respect to etiology of PAH. We hypothesized that patients with underlying congenital heart disease associated PAH (CHD-APAH) will have shorter hospital stay and mortality compared to patients with non-CHD PAH. In addition, we analyzed outcomes with respect to management strategies (BAE versus conservative management).

Methods

Most PAH patients followed at the Baylor Pulmonary Hypertension Center requiring hospitalization or outpatient procedures are admitted to The Methodist Hospital (TMH). All records were queried from 2001–2009 using discharge ICD-9 codes for hemoptysis (786.3) and primary pulmonary hypertension (416.0). Patients were excluded if pulmonary hypertension (PH) was not confirmed or PH diagnostic group was II, III, IV, or V. (Figure 1) Demographic, hemodynamic, procedural, laboratory and clinic data was reviewed.

Patients were divided into two groups based on etiology of PAH; CHD-APAH and non-CHD related PAH. Patients were also grouped according to the type of therapy received for hemoptysis; BAE versus conservative treatment group. Patients treated conservatively received standard non-surgical interventions including blood product transfusions and discontinuation of anticoagulant therapies. Patients in the BAE group received BAE using polyvinyl alcohol particles performed by an interventional radiologist. Additional data for the BAE group included number and location of bronchial arteries embolized. Patients were included in the conservative treatment group if they did not receive BAE for hemoptysis.

Data is presented as mean±standard deviation or median with interquartile range, where appropriate. Data was analyzed and compared using Student's unpaired t-test. Categorical

outcome data was analyzed using Fisher's exact test. Kaplan-Meier survival plots were analyzed using the log-rank test. A p value of <0.05 was considered significant. A Cox proportional hazard model was utilized to determine the effects of etiology and treatment type with respect to survival status accounting for a variable follow up period. Statistics were performed using SAS 9.2 (SAS Institute Inc., Cary, NC) software package.

Results

A total of 145 admissions from 116 PH patients were reviewed (Figure 1). Thirty-three patients with diagnostic group II pulmonary hypertension, 6 patients with diagnostic group III pulmonary hypertension and 11 patients with diagnostic group IV pulmonary hypertension were excluded. The remaining 45 patients were excluded for not having PH or not presenting with hemoptysis. Twenty-one PAH patients presenting with hemoptysis for 37 admissions were included in this study. Eight patients were classified as having CHD-APAH, which included 4 patients with Eisenmenger syndrome, 1 patient with a corrected shunt, 1 patient with transposition of the great arteries, 1 patient with congenital pulmonary artery stenosis and 1 patient with situs inversus.

Demographic and clinical data for each group is presented in Table 1. There were no significant differences in baseline demographics, hemodynamics and laboratory values between the two groups except for body mass index and hematocrit (Table 2). Hemodynamic data listed represents echocardiographic results obtained during the hospital admission. Comparing CHD-APAH patients with all other PAH patients, 28-day mortality (0% vs. 31%) and 1-year mortality (0% vs. 54%) were lower in the CHD-APAH patients. A statistically significant difference was found in the survival fraction for full follow up time when comparing CHD-APAH patients and non-CHD PAH patients ($p=0.02$) (Figure 2). Length of hospitalization between these two groups was not statistically different.

When comparing treatment modalities, 14 patients with 29 admissions were included the BAE therapy group, and 7 patients with 8 admissions were in the conservative group. The presenting amount of hemoptysis ranged from streaking to massive (>500 cc). Admission medications included warfarin in 14% of BAE patients (2/14) and 14% of conservatively treated patients (1/7). The patients who were on warfarin had a sub-therapeutic INR at the time of admission (<1.5) and warfarin was subsequently discontinued during and following hospitalization

One patient in the BAE group and three patients in the conservative group were not on PAH-specific therapy at the time of admission. These patients were either newly diagnosed with PAH or had not yet been referred to the Baylor PH Center.

Bronchial artery embolization was well tolerated and very successful in terminating hemoptysis. The procedure was successfully completed 96% (29 of 30) of the time (Figure 3). The single embolization failure was due to the inability to cannulate the femoral artery. An average of 2.4 ± 1.2 bronchial arteries were embolized per procedure. The particle size varied among each patient, and was selected according the expertise of the attending

interventional radiologist. There was recurrent hemoptysis in 43% of patients treated with BAE with a median time to re-bleed of approximately 3 months.

When comparing the two treatment groups, patients treated with BAE had non-significant shorter length of stays (4.0 ± 4.0 vs. 13.7 ± 22.5), and non-significant trends toward decreased 28-day and 1-year mortality rates. 28-day mortality was 14% (2 of 14) in the BAE group and 28.5% (2 of 7) in the conservatively managed group ($p=0.57$). 1-year mortality was 21% (3 of 14) in the BAE group and 57% (4 of 7) in the conservatively managed group ($p=0.16$). Survival curve and log rank values are illustrated in Figure 4. There was a 28-day mortality rate of 19% and a 1-year mortality rate of 33% for the combined groups.

The follow up time available for each patient was different with a median of 36 months (IQR 3.6 to 59.1 months). A statistically significant difference was not identified in the survival fraction for the full available follow up time between the two treatment groups ($p=0.31$) (Figure 4). A Cox proportional hazard model showed no significant effect of treatment type on survival status of patients adjusted for the follow up time differences ($p=0.35$). Excluding CHD-APAH patients from the analysis resulted in a combined 28-day mortality of 30% and 1-year mortality of 54% for both BAE and conservatively treated groups. A Cox proportional hazard model revealed no significant effect of treatment type (BAE or conservative treatment) on survival with CHD-APAH patients excluded ($p=0.28$).

Discussion

The median survival for patients with PAH was previously reported as 2.8 years.¹⁰ In the modern treatment era, median survival has been substantially prolonged.^{11,12} Historically, hemoptysis was relatively rare occurring more commonly in patients with congenital heart disease, and frequently reported as a terminal event.¹³ With greater longevity, the consequence of PAH includes the development of robust bronchial collaterals with their consequent risk of arterial bleeding and resulting hemoptysis.

There is a paucity of published literature regarding outcomes of hemoptysis in PAH patients. Unpublished data by Jais et al revealed 20 consecutive heritable or idiopathic PAH patients admitted with hemoptysis either treated conservatively ($n=8$) or with embolization ($n=12$). Mortality was 25% at 30 days for the BAE group and 63% at 30 days for the conservatively managed group.³ These results were not statistically different and very similar to our findings of a large absolute mortality differences between the two groups that did not meet statistical significance.

Our study cohort includes a relatively large proportion of CHD-APAH patients. These patients had a statistically significant lower rate of mortality following initial episode of hemoptysis when compared to all other subtypes of PAH. Hemoptysis is a common complaint in patients with Eisenmenger syndrome which does not appear to bestow increased short term mortality.^{5,14} For patients with non-Eisenmenger APAH, it is unknown if hemoptysis significantly increases mortality following the event.⁴

As previously described in the literature, BAE is highly effective in terminating bleeding. However, there is a high incidence of recurrent bleeding.^{4,7-9} Our study corroborates this

phenomenon with 33% of all patients treated with BAE re-presenting with hemoptysis. Despite therapy with BAE, there was still an increased risk of death at the 1-year follow up. Zylkowska et al reported a 50% cumulative survival rate at 3 month follow up, and Jais et al reported 36% survival rate at 1 year follow up in PAH patients with hemoptysis treated with BAE.³⁴ The follow up survival rates in our cohort are higher than previously reported. There was a 3 month survival rate of 76% and a 1 year survival rate of 67% in our patient population. Following exclusion of CHD-APAH patients from our full cohort, the 28 day survival rate of 70% and 1 year survival rate of 46% was still higher than previously reported literature. The difference may be accounted for by the heterogeneous nature of all subtypes of PAH included in this study. The patient population in the study by Zylkowska et al included a large amount of hereditary PAH patients with multiple episodes or re-bleeding. The one hereditary PAH patient in our study also had multiple re-bleeding episodes (n=9). Unidentified genetic factors inherent to hereditary PAH may predispose these patients to recurrent bleeding, including the possibility of unrecognized hereditary hemorrhagic telangiectasis. Given the high mortality rate, supportive care alone does not seem to be a viable option for therapy in this patient population.

The observed high mortality rate has prompted many PAH physicians to refer early for lung transplantation. Bronchial artery embolization as a temporizing measure for termination of acute hemoptysis with subsequent referral for lung transplantation seems to be an appropriate strategy suggested by some investigators. The increasing survival rates of lung transplantation in PAH patients also supports this strategy. Other options for hemoptysis therapy include surgical resection, flexible bronchoscopy and emergency lung transplantation. The cardiopulmonary complications innate to PAH predispose these patients to a very high risk for any emergency surgery. The frequent inability to localize bleeding on flexible bronchoscopy renders endobronchial therapies much less effective.⁴¹⁵ Emergency lung transplantation is not a realistic option due to logistics and a high failure rate in the acute situation.¹⁶ For patients with CHD-APAH, utilization of BAE seems to be the optimal therapy of choice, as the patients seem to have an inherent resilience to the mortality risk of hemoptysis.

Prostaglandins have both antiplatelet and antithrombotic properties, which could theoretically increase the chance of bleeding. In our cohort, 19% of patients were not on PAH specific therapies, 38% of patients were on non-prostaglandin treatments (ETRA or PDE-5 Inhibitor only) and 43% of the patients were on prostaglandin treatment. Prostaglandins were administered intravenously in the majority of the patients (8 of 9 patients), with the remaining patient on inhaled prostaglandin therapy. Patients with thrombocytopenia, abnormal liver function, abnormal coagulation profiles or significant renal dysfunction may also have a higher risk of bleeding.¹⁷ These laboratory abnormalities were essentially absent in our cohort.

Hemoptysis is the result of engorged bronchial arteries with abnormal collateral vessel formation and systemic arterial pressures similar to that described in patients with chronic thromboembolic pulmonary hypertension (CTEPH).¹⁸ Patients with CTEPH have been reported to have a high incidence of enlarged bronchial arteries and are also known to experience hemoptysis.² Pulmonary circulation is reduced at the level of the pulmonary

arteriole due to vascular remodeling, hypoxemic vasoconstriction and microthrombosis.¹⁹ This lack of blood flow gives rise to bronchial artery angiogenesis with subsequent temporal enlargement.²⁰ These vessels may rupture, due to elevated regional blood pressure, and leak into the tracheobronchial tree causing hemoptysis.²¹

Although no improvement in mortality was noted by performing BAE, it appears that the procedure is safe and effective for terminating hemoptysis. The BAE group fared no worse than those treated solely with conservative measures. With no excessive morbidity and a shorter length of stay noted, treatment with BAE seems to be a reasonable therapeutic measure. In addition, the excessive mortality related to hemoptysis may have been prevented due to the early intervention and treatment. However, a larger sample size would be necessary to confirm these findings.

Limitations for this study include a small sample size and the retrospective nature. In addition, this cohort does not include PAH patients seen in the emergency center who may have been discharged without admission. The PAH group had a higher BMI than the CHD-APAH group, which is unlikely to significantly confound the results given such a short period. The statistically significant higher hematocrit found in CHD-APAH patients could theoretically cause higher morbidity, however, this effect was not observed.²² The conservatively treated group was older than the BAE group, which could confound the unadjusted mortality results. One patient had mild renal dysfunction evidenced by a high BUN, which may increase propensity of bleeding through platelet dysfunction and may confound the results of the conservatively treated group. There were also fewer conservatively treated patients on PAH specific pharmacotherapy. The BAE group also had a higher proportion of CHD-APAH patients, although no statistically significant mortality differences were seen when these patients were excluded from analysis.

Conclusions

When comparing CHD-APAH patients with non-CHD PAH patients presenting with hemoptysis, statistically significant lower mortality rates were found. CHD-APAH seems to confer a survival advantage, independent of therapy utilized. Therapeutic choice did not influence length of stay nor mortality in our cohort. However, a small sample size may have confounded the results evidenced by a large absolute difference between the two treatment groups. With no worse morbidity and mortality in patients treated with BAE, it is reasonable to perform BAE for acute termination of hemoptysis in patients with PAH. Bronchial artery embolization is highly successful resulting in rapid cessation of hemoptysis with low complication rates. For patients with CHD-APAH, BAE is the ideal therapy for rapid resolution of bleeding. For non-CHD PAH, BAE will also effectively terminate hemoptysis. However, re-bleeding is common, and referral for transplant consideration is prudent following the initial hemoptysis episode.

Acknowledgments

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Figure 1.

All charts of patients admitted with hemoptysis and pulmonary hypertension were reviewed. The majority of patients that were excluded did not have PAH. Twenty-one patients were included in this cohort.

Figure 2.

Survival plot for full follow up period comparing CHD-APAH and non CHD-APAH group. Survival for CHD-APAH group was significant better than the non-CHD PAH group. One patient died in CHD-PAH group whereas there were eight deaths in non-CHD PAH group.

Figure 3.

Successful Bronchial Artery Embolization in a patient. Panel A. Diminutive second order vessels from the left internal mammary artery demonstrating blushing and dilatation (arrow). Panel B. None of the previously seen branches are visible after successful embolization to stasis.

Figure 4.

Survival plot as calculated for the full follow up period for both groups. Median follow up time for all patients was 36 months. The survival rate for the combined groups over the full follow up period is 52%.

Table 1

Demographic and Clinical Data

	CHD-APAH group	Non CHD-APAH group	p Value
Subjects, N	8	13	–
Age, years	38.1 ± 14.2	43.3 ± 15.2	0.51
Female, n (%)	5 (63%)	9 (69%)	–
Body Mass Index, kg/m ²	23.9 ± 4.8	31.9 ± 10.7	0.04*
Race, n (%)			–
White	4 (50%)	9 (69%)	
Black	2 (25%)	2 (15%)	
Hispanic	2 (25%)	2 (15%)	
Treatment type, BAE (%)	5 (63%)	8 (62%)	
Medication at Initial Admission			
Warfarin	1 (13%)	1 (8%)	
Digoxin	8 (100%)	8 (62%)	
Prostacyclin	2 (25%)	7 (54%)	
Endothelin Receptor Antagonist	4 (50%)	2 (15%)	
Phosphodiesterase-5 Inhibitor	3 (38%)	3 (23%)	

Data is presented as mean ± standard deviation. A p <0.05 was considered statistically significant.

Table 2

Hemodynamic and Laboratory Values at Admission

	CHD-APAH group	Non CHD-APAH group	p Value
Subjects, N	8	13	–
Hemodynamics			
Pulmonary Arterial Systolic Pressure, mmHg	84 ± 23	93 ± 28	0.44
Cardiac Index, L/min/m ²	3.13 ± 1.77	2.19 ± 0.60	0.31
Right Atrial Pressure, mmHg	9.2 ± 5.2	10.1 ± 6.0	0.74
Laboratory Values			
Platelet, 10 ³ /μL	188 ± 87	155 ± 99	0.43
INR	1.2 ± 0.2	1.3 ± 0.3	0.79
Blood Urea Nitrogen, mg/dL	15.6 ± 12.9	25.6 ± 16.3	0.15
Creatinine, mg/dL	0.9 ± 0.2	1.2 ± 0.4	0.06
Hematocrit, %	50.8 ± 5.9	39.3 ± 6.8	0.01*
Alkaline Phosphatase, U/L	110 ± 27	101 ± 39.2	0.61
ALT, U/L	21.6 ± 15.0	29.0 ± 22.5	0.45
AST, U/L	26.6 ± 9.9	27.4 ± 18.1	0.90
Albumin, g/dL	4.4 ± 0.2	3.9 ± 0.7	0.07

Data is presented as mean ± standard deviation. A p < 0.05 was considered statistically significant.

Table 3

Bronchial Artery Embolization Group

Pt.	Age, yrs	Gender	Etiology	WHO FC	6MWD, m	Follow up, mths	Duration of Illness, mths	Re-bleed Episodes	Outcome
1	46	F	EIS	III	–	29	NK	–	Alive
2	65	F	EIS	III	368	39	631	–	Alive
3	41	M	EIS	II	440	59	NK	–	Alive
4	39	F	CPAS	III	93	38	254	3	Alive
5	24	M	IPAH	–	–	0	NK	–	Expired
6	55	F	CTD	II	488	0	35	–	Expired
7	53	F	IPAH	III	117	4	20	–	Expired
8	35	F	IPAH	III	360	21	32	1	Expired
9	42	F	IPAH	III	290	36	46	–	Alive
10	20	F	SI	III	180	61	39	1	Alive
11	21	F	COR	II	329	56	233	1	Expired
12	20	F	CTD	III	128	36	New Dx	–	Alive
13	29	M	HPAH	III	400	78	32	8	Alive
14	42	F	IPAH	II	495	69	77	1	Alive

M, male; F, female; WHO FC, WHO functional class; 6MWD, six-minute walk distance; EIS, Eisenmenger Syndrome; CPAS, congenital pulmonary artery stenosis; SI, situs inversus; COR, corrected shunt; CTD, connective tissue associated PAH; IPAH, idiopathic PAH; HPAH, hereditary PAH; m, meters; NK, not known.

Table 4

Conservative Treatment Group

Pt.	Age, yrs	Gender	Etiology	WHO Class	6MWD, m	Follow up, mths	Duration of Illness, mths	Re-bleed Episodes	Outcome
15	76	Female	CTD	III	–	1	3	–	Expired
16	58	Female	IPAH	III	45	3	8	–	Expired
17	40	Male	TGA	–	–	36	NK	–	Alive
18	39	Female	IPAH	–	–	102	134	–	Expired
19	49	Male	PPHTN	III	–	8	12	–	Expired
20	39	Male	EIS	III	430	103	NK	1	Alive
21	41	Female	IPAH	–	167	0	24	–	Expired

M, male; F, female; WHO FC, WHO functional class; 6MWD, six-minute walk distance; TGA, transposition of the great arteries; EIS, Eisenmenger syndrome; CTD, connective tissue associated PAH; IPAH, idiopathic PAH; PPHTN, porto-pulmonary hypertension; m, meters; NK, not known.