Clinical Features and Management of Chronic Thromboembolic Pulmonary Hypertension in Thai

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Background: *The incidence and clinical features of chronic thromboembolic pulmonary hypertension (CTEPH) in Thailand are unknown. This may be due to the difficulties in diagnosis, poor disease recognition and non-specific presenting symptoms and signs. With the new revolution of diagnostic tools and advanced knowledge about this condition, CTEPH is now discovered much more often.*
Study design: *Retrospective cross-sectional study*
Objective: *To evaluate common risk factors, symptoms, signs, commonly used investigations, treatment and results of treatment in Thai patients with CTEPH.*
Material and Method: *All patients with confirmed CTEPH diagnoses were selected for analysis in this study. We evaluated characteristics of patients with CTEPH at Siriraj hospital. All CTEPH patients' data included demographics, symptoms, signs, investigations, treatments, outcome and risk factors such as malignancy; previous deep vein thrombosis, immobilization, and congenital thrombophilia were recorded.*
Statistical analysis: *The descriptive statistics were used in this study. Categorical data and continuous data presented in term of percent and mean with standard deviation respectively.*
Results: *fourteen patients had confirmed diagnoses of CTEPH, five patients were male and 9 were female. The age of patients varied from 28 to 79 years old. Mean age was 55 years old. The most common presenting symptoms and signs were dyspnea (100%), followed by leg edema (78.6%), tachypnea (71.4%) and accentuation of pulmonic component of second heart sound (loud P_2) (57.1%). At the time of diagnosis, four out of 14 patients were in NYHA class II, ten were in NYHA class III, and none of them was in NYHA class IV. Idiopathic CTEPH was found in 54.1% of patients. Congenital and acquired thrombophilia were the frequent risk factors found in our series. The most frequent radiographic abnormalities included enlarged pulmonary trunk, right atrium/right ventricle (RA/RV) dilatation, and cardiomegaly followed by prominent pulmonary trunk. Echocardiography findings were elevated right ventricular systolic pressure (RVSP); ranges varied from 60 mmHg to 137 mmHg (mean was 107.48 mmHg). The most common finding of arterial blood gas in APE patients was respiratory alkalosis with hypoxemia. The most frequent diagnostic tool used was ventilation-perfusion lung scan. Treatment of CTEPH patients was by anticoagulant, pulmonary thromboendarterectomy and inferior vena cava filter insertion. The success rate of surgical thromboendarterectomy in our institute was about 80%*
Conclusion: *Characteristics of CTEPH patients are unique and different from acute pulmonary embolism in aspects of clinical presentations, risk factors, investigation, common findings, treatment and outcome.*

Keywords: *Clinical features, CTEPH, Pulmonary embolism, Thromboendarterectomy*

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Natural history of acute pulmonary embolism (APE) is either complete resolution with restoration of normal pulmonary hemodynamics, gas exchange and exercise tolerance or partial resolution leaving minimal residual causing long term change in pulmonary hemodynamics and pulmonary arterial hypertension with deterioration of right ventricular function which is called chronic thromboembolic pulmonary hypertension (CTEPH)[1-4]. CTEPH is a condition which occurs in a

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minority of patients after APE[5]. The exact incidence of CTEPH is unknown, the estimated incidence is 0.1 to 0.5 percent in survivors of acute pulmonary emboli[1].

Clinical manifestations of CTEPH are the same as in other forms of pulmonary hypertension. The most common manifestation are progressive exertional dyspnea and exercise intolerance[1]. The findings on physical examination are an accentuation of the pulmonic component of the second heart sound (P_2), and other signs of pulmonary hypertension.

Initial work up for CTEPH consists of a chest radiograph (CXR), electrocardiograph (EKG) and arterial blood gas analysis (ABG) which often show evidence of pulmonary arterial hypertension. Evidence of pulmonary arterial hypertension is seen by a large pulmonary arterial shadow on the CXR, right axis deviation and S wave in lead I, Q wave and T inversion in lead III from the EKG and mild hypoxemia with respiratory alkalosis from ABG). These findings are nonspecific. Echocardiography (echo) is usually performed to confirm and assess the degree of pulmonary hypertension[6]. The definite diagnosis of CTEPH is usually obtained by a ventilation-perfusion lung scan (V/Q), computerized tomography of the chest with contrast (CTA) or pulmonary angiography (PAgram) and, sometimes, pulmonary angioscopy[7,8]. The only curative treatment of CTEPH is pulmonary thromboendarterectomy (PTA). A successful treatment is dependent on the position of the clots in the pulmonary vasculature, functional status of the patients and patients' co-morbid diseases[9].

The incidence and clinical features of CTEPH in Thailand is unknown. This may be due to difficulties in diagnosis, poor disease recognition or non-specific presenting symptoms and signs of this condition. There were only a few sporadic case reports of CTEPH in Thailand. With the new revolution of diagnostic tools and advanced knowledge about this condition, more cases of CTEPH were discovered. The purpose of this study was to assess the prevalence, risk factors, clinical manifestation and management of the patients with CTEPH who presented as pulmonary embolism at Siriraj Hospital.

Material and Method

Patient selection

All adult (age of 15 year-old and over) patients with ICD coding of I26.0 (pulmonary embolism with acute cor pulmonale) and I26.9 (pulmonary embolism without acute cor pulmonale) who were admitted to Siriraj Hospital, Bangkok, Thailand between January 2001 and October 2005 were recruited in the study. The medical records were reviewed. All patients with confirmed CTEPH were included in the study.

The diagnostic criteria for CTEPH were patients with pulmonary hypertension plus the abnormal findings from one of these following diagnostic tests

1. High probability V/Q
2. Organized eccentric clot in pulmonary vasculature from CTA with mosaic pattern in lung parenchyma
3. Abnormal PAgram such as pouch defect, acute tapering of vascular lumen, roughening or pitting of the intimal surface, bands, webs traversing the vascular lumen, pitted masses of chronic embolic material within the lumen, and partial recanalization[7]

The data included demographics, baseline characteristics, symptoms, signs, risk factors (such as malignancy, previous deep venous thrombosis (DVT), congenital thrombophilia, immobilization, oral contraceptives, trauma and previous surgery), New York Heart Association (NYHA) functional class, investigations, treatment and outcome were recorded. The study protocol was approved by Siriraj Hospital Ethics Committee.

Statistical analysis

Descriptive statistics were used in this study. Categorical data and continuous data were presented in terms of percent and mean with standard deviation respectively.

Results

One hundred and seventy-two patients with ICD coding of 126.0 (pulmonary embolism with acute cor pulmonale) and 126.9 (pulmonary embolism without acute cor pulmonale) were identified. Of these 172 patients, 71 patients had acute pulmonary embolism while 14 patients met our diagnostic criteria for CTEPH. The rest had other diagnoses such as pulmonary hypertension, heart failure, hypoxia, etc and did not meet the specific diagnostic criteria for acute or chronic pulmonary embolism.

Patient's characteristics and risk factors

Of the 14 patients with CTEPH, five patients were male, 9 patients were female with the mean age of 55 years old (ranging from 28 to 79 years old). Two patients had previous DVT, 2 had congenital thrombophillia (one of them had protein C deficiency, one had protein S deficiency), 2 had acquired thrombophillia (antiphospholipid syndrome) and none

of the patients had the history of malignancy. Eight out of 14 patients with CTEPH had no risk factors for DVT or APE (Table 1).

Signs and symptoms

At the time of diagnosis, four out of 14 patients were in NYHA class II, ten were in NYHA class III, and none of them were in NYHA class IV. The most frequent presenting symptoms was dyspnea which was found in all the patients (100%), followed by leg edema (78.6%), cough (35.7%) and chest pain (21.4%). Syncope occurred in only one case (7.1%). The most common sign was tachypnea which was found in 10 out of 14 patients (71.4%), followed by an accentuated second heart sound (loud P_2) which represent pulmonary arterial hypertension. None had a continuous murmur over the chest (Table 2).

Table 1. Risk factors of patients with chronic thromboembolic pulmonary hypertension

Risk factors (n = 14)	Frequency (%)
Immobilization	0
Trauma	0
Surgery	0
Obesity	0
Pregnancy	0
Hormonal treatment	0
Previous DVT	2 (14.3)
Malignancy	0
Congenital thrombophilia	2 (14.3)
Antiphospholipid syndrome	2 (14.3)
None of above	8 (54.1)

n = Total number of patients

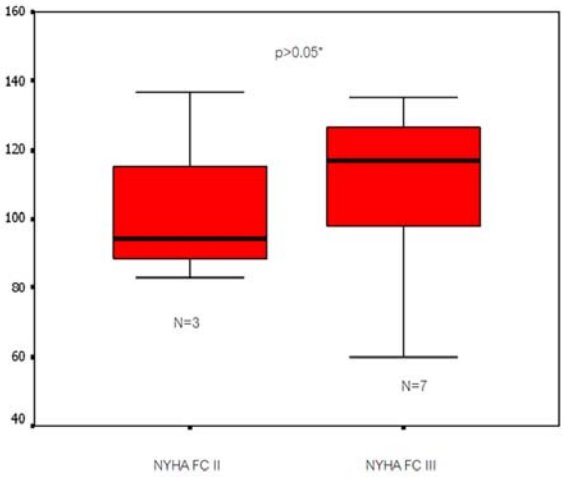
Table 2. Symptoms and signs of patients with chronic thromboembolic pulmonary hypertension

Symptoms n = 14	Frequency (%)	Signs n = 14	Frequency (%)
Dyspnea	14 (100)	Tachycardia	8 (57.1)
Chest pain	3 (21.4)	Tachypnea	10 (71.4)
Palpitation	0	Hypotension	0
Cough	5 (35.7)	Cardiac arrest	0
Hemoptysis	1 (7.1)	Rhonchi	1 (7.1)
Cyanosis	1 (7.1)	Crepitation	1 (7.1)
Leg swelling	11 (78.6)	S_4	2 (14.3)
Syncope	1 (7.1)	Fever	1 (7.1)
		Loud P_2	8 (57.1)
		Continuous murmur	0

n = Total number of patients

Investigation

Of the 14 patients with a diagnosis of CTEPH, 4 of 14 patients were diagnosed via V/Q, 2 by CTA and none by PA gram alone. Diagnosis was made by both V/Q and CTA in 5 patients, by both V/Q and PAgram in 1 patient and by all 3 tests in 2 patients. Almost all the patient with CTEPH had an echocardiography finding which demonstrated right atrium and right ventricular dilatation, tricuspid regurgitation, dilatation of pulmonary trunk or pulmonary artery, and leftward displacement of the interventricular septum with normal left ventricular ejection fraction (LVEF). Only one had a visible clot in pulmonary trunk, and four of them demonstrated ventricular chamber enlargement by echocardiography. The mean estimated right ventricular systolic pressure (RVSP) obtained from echocardiography of 10 out of 14 patients was 107.48 $\pm$ 26.99 mmHg. Of the two patients who had a PAgram, the mean pulmonary-artery pressure (mean PAP) was 55 mmHg and 65 mmHg, systolic pulmonary artery pressure (SPAP) was 95mmHg and 94 mmHg, respectively. PA gram findings suggested proximal organized thrombi in 1 out of 3 patients and a distal lesion in 2 out of 3 patients. Estimated RVSP of each NYHA function class were shown in Fig. 1. All of the patients with CTEPH



RVSP = Right ventricular systolic pressure
NYHA FC = New York heart association function class
*p > 0.05 = non statistically significant (Mann-Whitney U test)

Fig. 1 Estimated RVSP in 2 NYHA functional class groups

had RVSP or PA systolic pressure over 60 mmHg. One patient who had syncope had an RVSP of 83 mmHg.

CTA finding was varying, mosaic attenuation of the pulmonary parenchyma and eccentric organized thrombi positioned within the central pulmonary arteries were seen in three patients who had CTA.

The different EKG findings in CTEPH are seen in Table 3. Two of the most common abnormalities were $S_1Q_3T_3$ (41.7%), followed by right axis deviation (33.3%). CXR findings included enlarged pulmonary trunk, right atrium and right ventricle dilatation, and cardiomegaly. Seven patients had doppler ultrasonography done on their legs. Interestingly, only 28.6% of patients showed evidence of DVT. Serum D-Dimer level measurement using Latex agglutination technique showed the level below 500 IU in four out of 9 patients (44.4%) and the level of 500 IU and over in 5 out of 9 patients (55.6%).

Treatment and outcome

Unfractionated heparin was used in 6 out of 14 CTEPH cases (42.9%) and low molecular weight heparin (LMWH) was used in 4 out of 14 CTEPH patients (28.6%). One patient received thrombolytic agents due to an acute large pulmonary emboli superimposed CTEPH. Five patients (35.7%) underwent pulmonary thromboendarterectomy and IVC filters were placed in 6 patients (42.9%), as in Table 4.One patient died from CTEPH in the postoperative period. Four patients had marked improvement after thromboendarterectomy, their NYHA improved from class III to class I or class II. The remaining patients who did not have other interventions except oral anticoagulants remained stable.

Discussion

Data from North American studies showed that CTEPH, a distinct but serious complication of acute pulmonary emboli, is relatively rare. It occurs approximately 0.1-0.5 percent of patients with pulmonary emboli who survived. According to our study, we found 14 patients at Siriraj Hospital between January 2001 and October 2005 with CTEPH. This may imply that the estimated number of APE cases could be 140-700 cases per 5 years or 30-180 cases per year which is much more than we expected. Most of our patients with CTEPH presented late in the course of the disease. Patients with thromboembolic disease may remain asymptomatic for months or years before the first symptom is noticed. Most of our patients were in NYHA class III by the time of the diagnosis. The most common presenting symptoms were progressive exertional dyspnea and exercise intolerance which were the same as in previous reports[1,13]. The common physical signs are the signs of right sided heart failure secondary from pulmonary hypertension such as legs edema, tachypnea and tachycardia and loud P2. Syncope, which is thought to be the result of severe pulmonary hypertension and the inability of a compromised right ventricle to introduce adequate cardiac output, was found in only one patient in our study group. All of our CTEPH patients had RVSP of over 60 mmHg which is higher than the average RVSP of patients with acute pulmonary embolism. NYHA functional class of CTEPH patients were primarily in class III, the higher functional class seemed to be correlate with the higher RVSP.

Congenital and acquired thrombophilia were more prevalent in our CTEPH patients (28.6%) whereas none of these CTEPH patients had malignancies, which were the most common risk factors in patients with APE or DVT in our previous study[10]. Congenital thrombophilia may affect the balance of coagulation and thrombolytic process, which may lead to the long-term development of organized, non-resolving thrombi in pulmonary vasculature causing CTEPH. Although, malignancy with hypercoagulable state may frequently cause DVT and APE, patients with malignancy and APE may not live long enough to develop CTEPH.

Table 3. EKG findings in chronic thromboembolic pulmonary hypertension patients

EKG findings (n = 14)	Frequency (%)
Sinus tachycardia	2 (16.7)
S1Q3T3	5 (41.7)
Right axis deviation	4 (33.3)
Non-specific ST-T change	3 (21.4)

n = Total number of patients

Table 4. Treatment of the patients with chronic thromboembolic pulmonary hypertension

Treatments (n = 14)	Frequency (%)
Unfractionated heparin	6 (42.9)
LMWH	4 (28.6)
Thrombolytic therapy	1 (7.1)
IVC filter	6 (42.9)
Pulmonary thromboendarterectomy	5 (35.7)

n = Total number of patients

Initial laboratory tests such as CXR and EKG are all nonspecific and similar to other cases of unexplained pulmonary hypertension. Duplex scanning of the legs revealed evidence of prior venous thrombosis in 32.5% of patients with CTEPH in our study, which is comparable to the previous reports[11,12]. Transthoracic echocardiogram demonstrated variable degrees of right atrial and right ventricular enlargement, tricuspid regurgitation, a leftward displacement of the interventricular septum which are similar findings as in other causes of pulmonary arterial hypertension. V/Q scan demonstrated high probability in almost all patients. Approximately 50% of CTEPH cases were diagnosed by using CTA. However, PAgram is still an essential tool not only to confirm the diagnosis of CTEPH by revealing the characteristic of clots and degree of pulmonary hypertension but also the extent and location of the disease, which determines the operability. CTEPH patients with severe pulmonary hypertension and proximal vascular lesions may be candidates for surgical thromboendarterectomy. Almost all patients who underwent pulmonary thromboendarterectomy in our institution had PAgram done prior the surgery.

All of our patients received either unfractionated heparin or LMWH initially and eventually received oral warfarin for life. A thrombolytic agent was not routinely used in CTEPH because it does not affect the chronic organized thrombi even though one of our fourteen patients received intralesional thrombolytic injection due to demonstration of intraluminal large clots in right and left pulmonary artery during PAgram. Later on, that patient had surgical thromboendarterectomy.

Surgical thromboendarterectomy is a complex, complicated, sophisticated procedure that can be done in only a few centers around the world where highly experienced cardiothoracic surgeons and teams are present. Five of our CTEPH patients with severe pulmonary hypertension (NYHA class III) with proximal lesions underwent pulmonary thromboendarterectomy. Four patients had successful surgeries but one patient died postoperatively due to arrhythmia and persistent right heart failure. Estimated surgical mortality was 20% in our institution, which is comparable to other institutions (around 5-20%)[13-15]. Nine CTEPH patients with distal clots did not have thromboendarterectomy. Inferior vena cava filers (IVC filter) were placed in all 5 patients who underwent thromboendarterectomy and in another one CTEPH with distal thrombi who was not a surgical candidate due to a past history of previous intracranial hemorrhage and seizure.

Only two of fourteen patients died. One of them died postoperatively. Another one had acute pulmonary emboli superimposed CTEPH with fatal right ventricular failure. Four patients who survived thromboendarterectomy had much improved NYHA class. Remaining patients were stable with anticoagulation.

Conclusion

CTEPH, a unique type of pulmonary hypertension, is an unusual but curable complication of acute pulmonary embolism. The true prevalence of this condition is unknown. In our study we found 14 CTEPH cases within a 5 year period. The clinical manifestations, baseline characteristic, symptoms and signs of these patients were not different from previous reports in European countries. Interestingly, we observed different congenital and acquired thrombophilia factors in up to 20% of our CTEPH cases. Echocardiogram, V/Q, CTA and PAgram play important roles in the diagnosis, evaluation and determination of treatment options in these cases. Thromboendarterectomy is still a curative treatment in surgically accessible CTEPH with severe pulmonary hypertension. In our institute, the successful thromboendarterectomy rate was about 80%.

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ลักษณะทางคลินิกและการรักษาภาวะความดันหลอดเลือดแดงปอดสูงจากลิ่มเลือดอุดตันหลอดเลือดแดงปอดเรื้อรังในไทย

สุรีย์ สมประดีกุล, มนต์สวรรค์ มินิพันธ์

***บทนำ**: ภาวะความดันหลอดเลือดแดงปอดสูงจากลิ่มเลือดอุดตันหลอดเลือดแดงปอดเรื้อรัง (CTEPH) แม้จะเป็นภาวะที่พบได้ไม่บ่อย แต่มักมีความรุนแรงทำให้เกิดความพิการ และอัตราการตายจากหัวใจด้านขวาวายได้สูง โดยสมมุติฐานของการเกิดภาวะนี้ น่าจะเป็นผลตามมาจากการเกิดภาวะลิ่มเลือดอุดตันหลอดเลือดแดงปอดเฉียบพลัน ซึ่งมีการสลายของลิ่มเลือดไม่สมบูรณ์เกิดการอุดตันหลอดเลือดแดงปอดถาวร อัตราการเกิดภาวะนี้ในประเทศไทยไม่ทราบแน่ชัด*

***ลักษณะการศึกษา**: Retrospective cross-sectional study*

***วัตถุประสงค์**: เพื่อศึกษาลักษณะทางคลินิก ปัจจัยเสี่ยง วิธีการวินิจฉัย การรักษาและผลการรักษาภาวะนี้ในไทย*

***วัสดุและวิธีการ**: ทบทวนเวชระเบียนโดยเลือกผู้ป่วยที่ได้รับการวินิจฉัย เป็นลิ่มเลือดอุดตันหลอดเลือดแดงปอดเฉียบพลัน ที่เข้ารับการตรวจ และรักษาที่โรงพยาบาลศิริราชในช่วงเดือน มกราคม พ.ศ. 2543 ถึง ตุลาคม พ.ศ. 2548 โดยเกณฑ์การวินิจฉัย CTEPH คือ มีภาวะความดันหลอดเลือดแดงปอดสูง และมี ventilation/perfusion lung scan (V/Q scan) หรือ CT angiogram หรือ pulmonary angiogram (PA gram) ผิดปกติซึ่งเข้าได้กับภาวะ CTEPH*

***ผลการศึกษา**: พบผู้ป่วยที่ได้รับการวินิจฉัยเป็น CTEPH 14 ราย เป็นชาย 5 ราย เป็นหญิง 9 ราย อายุเฉลี่ย 55 ปี อาการที่พบบ่อยสุดคืออาการเหนื่อย (100%) รองลงไปคือขาบวม (78.6%) ความสามารถในการออกกำลังของผู้ป่วยส่วนใหญ่อยู่ใน New York Heart Association functional class III ส่วนใหญ่ของผู้ป่วยไม่พบปัจจัยเสี่ยงของลิ่มเลือดอุดตัน แต่พบความเสี่ยงมีลิ่มเลือดเกิดง่าย (congenital and acquired thrombophilia) มากถึงร้อยละ20 ภาพรังสีทรวงอกพบมีเงาหัวใจและเส้นเลือดแดงปอดโต การตรวจอัลตราซาวน์หัวใจพบมีความดันหัวใจช่องขวาสูงมากกว่า 60 มิลลิเมตรปรอทในผู้ป่วยทุกราย การสืบค้นที่ใช่ถี่มากสุดคือ V/Q scan แต่ในรายที่ได้รับการผ่าตัดเกือบทุกรายได้รับการตรวจฉีดสีในเส้นเลือดแดงปอด (PA gram) ก่อนผ่าตัด การรักษาประกอบด้วยการให้ยากันเลือดแข็งตัว การใส่ตะแกรงกรองลิ่มเลือดที่เส้นเลือดดำใหญ่บริเวณช่องท้องและที่สำคัญที่สุดคือการผ่าตัดลอกลิ่มเลือดจากเส้นเลือดแดงปอด (pulmonary thromboendarterectomy) ซึ่งถ้าประสบผลดี ภาวะความดันหลอดเลือดแดงปอดสูงจะทุเลาจนเป็นปกติหรือเกือบปกติ อัตราการผ่าตัดสำเร็จในผู้ป่วยกลุ่มนี้ในไทยประมาณร้อยละ 80*

***สรุป**: ภาวะ CTEPH พบได้ไม่บ่อยแต่มักมีความรุนแรงและมีอัตราตายสูง ลักษณะทางคลินิก การวินิจฉัย การรักษานั้นแตกต่างจากภาวะลิ่มเลือดอุดตันหลอดเลือดแดงปอดเฉียบพลันอย่างมาก โดยเฉพาะในด้านปัจจัยเสี่ยงของการเกิดและค่าความดันหลอดเลือดแดงปอดเฉลี่ย ซึ่งใน CTEPH นั้นค่าความดันหลอดเลือดแดงปอดเฉลี่ยมีค่าสูงมาก และภาวะ CTEPH มีการรักษาให้หายขาดได้โดยการผ่าตัด แต่มีข้อจำกัดในด้านอัตราการตายจากการผ่าตัดและสภาพร่างกายของผู้ป่วย*