

# PHLEGMASIA ALBA & CERULEA DOLENS "A CHALLENGE TO THE VASCULAR SURGEON"

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## ABSTRACT

**Phlegmasia Alba and Cerulea Dolens with venous gangrene results from acute massive venous thrombosis and obstruction of venous drainage of an extremity, usually the lower limb. It is a rare condition. It results in venous thrombo-embolism, leading to pulmonary embolism in 50% of untreated patients.**

**We received five cases of Phlegmasia Alba / Cerulea dolens in our surgical unit during 2002.**

**The diagnosis was based on clinical examination and Doppler study. Two patients were already suffering from venous gangrene of the foot. Embolectomy was done using the Fogarty technique. Two patients ended up with below knee amputations. One patient died seven days postoperatively.**

**All patients, when received were started on heparin therapy, which was continued perioperatively. Oral anticoagulants were continued till 6 months postoperatively. These patients are still being followed up.**

**PAD = Phlegmasia Alba Dolens**  
**PCD = Phlegmasia Cerulea Dolens**  
**PACD = Phlegmasia Alba & Cerulea Dolens**  
**DVT = Deep Venous Thrombosis**

## INTRODUCTION

Phlegmasia Alba Dolens & Cerulea Dolens with venous gangrene is a relatively uncommon but a surgically significant condition.

It is characterized by thrombosis of major venous channels of the extremities leading to decreased or complete cessation of venous drainage.

In the 16th Century Fabricius Hildanus was the first to describe the clinical syndrome which is now called PCD. In 1938, Gregoire made an outstanding description of the disease and used the term PCD to differentiate ischaemia associated massive venous thrombosis from Phlegmasia Alba Dolens which describes fulminant venous thrombosis without ischaemia. In 1939, Leriche and Gessendorfer performed the first thrombectomy in cases of PCD.

In the USA, 6,00,000 cases of thrombo-embolism are reported annually. PAD, PCD & venous gangrene can occur at any age but are more common during 5th and 6th decades of life and are more common in females.

## PATIENTS & METHODS

In this study, 5 patients of DVT with PACD were included. All these patients were referred from the medical units. A detailed history was taken and examination performed. Duplex ultrasonography and other investigations like including a complete blood count and clotting screen were done.

## RESULTS & DISCUSSION

Four (80%) of these patients were female, one (20%) was male. Two (40%) patients were less than 40 years of age, three (60%) patients were more than 40 years of age. In all these cases PACD & venous gangrene were secondary to DVT of lower limb.

In 4 patients (80%), the left leg was involved while in 1 patient (20%), the right leg was involved. Three (60%) patients had features of

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PCD at the time of presentation i.e painful, swollen and cyanosed leg, while 2 (40%) patients had features of impending venous gangrene at the time of presentation, in the shape of blebs and bullae.

Two (40%) patients had clinical manifestations of shock which was secondary to significant fluid loss in the affected limb.

Two (40%) patients had history of recent childbirth. The single male patient was suffering with carcinoma of the prostate gland. Of the remaining two patients, one was a known diabetic with a history of poor control of diabetes who was admitted for management of diabetic ketoacidosis when developed this complaint. The remaining patient had no obvious pre-disposing factors except obesity.

**CAUSES OF PA / CD**  
TABLE 1

S.No.	Cause	No. Of Patients %
1	Recent Delivery	2 (40%)
2	Carcinoma Prostate	1 (20%)
3	Diabetes	1 (20%)
4	Unknown	1 (20%)

A Provisional diagnosis was made in all the patients on the basis of history and examination. The diagnosis was confirmed with the help of duplex scanning in all 5 patients. Venography could not be performed in these patients as this facility was not available.

Medical therapy was started in all these patients. This included injection of a bolus dose of heparin (80-100 units/kg body weight) followed by a continuous infusion at 15-18 units/kg/hr. The dose of heparin was titrated to the APTT of the patient (which was kept 2.0-2.5 times of control). The platelet count was monitored for early detection of heparin induced thrombocytopenia. Intra Venous fluids were

given to patients who were in shock. Surgical intervention (thrombectomy) was performed in all the five patients due to failure of medical therapy. In 2 (40%) patients the abdominal approach was used, while in remaining 3 (60%) the femoral approach was used as the patients were not fit for General Anaesthesia. Two (40%) patients who had impending gangrene at the time of admission later underwent below knee amputation, as the thrombectomy could not revert the ischaemic changes. One (20%) patient died seven days after thrombectomy. Two (40%) patients recovered completely and were discharge on oral anticoagulant therapy (Table-2)

This study showed that PACD is a less common, but serious, condition. It is associated with high morbidity and mortality. In our series, all 5 patients were referred from medical unit where they remained on conservative treatment for 2-3 days prior to referral for surgery.

**OUTCOME OF PATIENTS WITH PACD**  
TABLE 2

S.No.	Outcome	No. Of Patients %
1	Below knee amputation	2 (40%)
2	Complete recovery	1 (20%)
3	Mortality	1 (20%)

### CONCLUSIONS:

The most important fact in the management of such patients is timely diagnosis and early referral for surgical intervention once medical therapy fails, if morbidity and mortality are to be reduced.

Moreover, creating awareness and educating our health workers may play an important role in the prevention of this grave condition.

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