

# Purpura Fulminans in Childhood Meningococcal Septicaemia

## Predictors of Purpura Fulminans and Amputations

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### Introduction

Purpura fulminans (PF) is a disorder characterised by progressive cutaneous haemorrhage and necrosis as a result of dermal vascular thrombosis and disseminated intravascular coagulation (DIC). This leads to localised thrombosis giving rise to a patchy appearance whilst leading to a systemic coagulopathy. It is a devastating complication of meningococcal septicaemia, affecting 15-25% of patients and can result in widespread skin loss, amputations and late functional sequelae.<sup>1-4</sup>



Currently there is no early intervention proven to reduce progression of PF and reduce amputations. The use of fasciotomies is controversial, whilst the use of tPA has been associated with complications such as intra-cerebral bleeding.<sup>1-6</sup>

### Aims

- To audit all cases of meningococcal septicaemia and PF in Alder Hey Children's Hospital, Liverpool since 2007
- To ascertain whether there are any early factors or signs that may assist in early diagnosis of PF or predict the likelihood of amputation
- To compare our surgical management and outcomes of PF to those in published literature

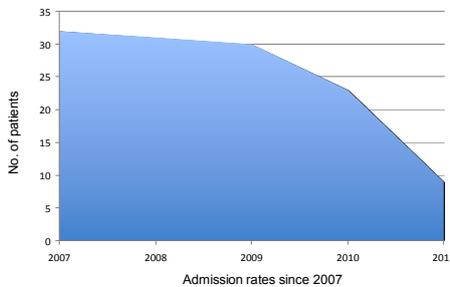
### Methods

A retrospective case note review of all patients admitted with suspected meningococcal septicaemia from 1<sup>st</sup> January 2007 to 31<sup>st</sup> May 2011 was undertaken. Electronic patient care records and laboratory data were retrieved to identify those patients who developed PF. PF was defined as any patient admitted to PICU who also had any area of skin necrosis. Patient demographics were recorded. Surgical interventions including amputations were also noted. Data was also recorded relating to investigations performed on admission including INR, APTT and platelet count.

Statistical analysis using t-tests were performed to identify significant differences in patients who developed PF and who had amputations.

### Results

During the study period, 125 patients were identified with suspected meningococcal sepsis. The mean age was 3 years 4 months (range 1 month 24 days to 17 years). The figure below illustrates the rate of admission over the last four and a half years.



#### Mortality

Overall, 4 patients died (2 in 2010, 1 in 2009 and 1 in 2007). Three patients died within 1 day of admission whilst the other died 4 days after admission. The overall mortality rate was 3.2%.

#### Purpura fulminans

We identified 15 patients (12%) who developed PF (6 males and 9 females). The mean age was 4 years 4 months (range 8 months to 17 years). All patients in this group had microbiological evidence of meningococcus Group B.

#### Surgical interventions

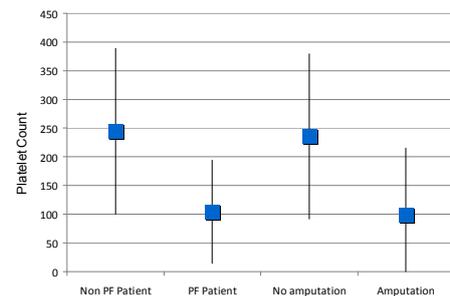
Seven patients with PF were treated conservatively. Eight patients required a form of surgical intervention. One patient only required debridement of wounds whilst 7 (47%) required an amputation of some degree. Five (33%) required at least one major limb amputation.

#### Predictors of PF and Amputation

We identified that admission platelet count and clotting tests may be able to predict development of PF and subsequent amputation.

#### Platelet Count

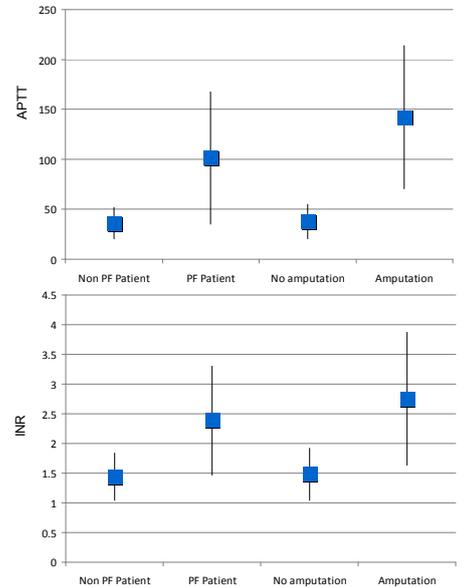
The figure below illustrates the differences in admission platelet counts between patients developing PF and those who did not. They also show differences between patients needing amputations and those who did not. The mean platelet count in a PF patient was 104.1 compared to 244.3 in a non PF patient ( $p < 0.005$ ). Similarly, in patients undergoing amputations the count was 98.7 compared to 144.2 ( $p = 0.02$ ).



#### Clotting Profile

The figures below illustrates the differences in admission APTT and INR between PF and non-PF patients. They also show differences in patients who needed amputations and those who did not.

Both APTT and INR were statistically significantly higher in patients developing PF ( $p < 0.0005$ ). They were also significantly higher in patients undergoing amputations ( $p < 0.05$ ).



### Discussion

Our mortality rate lies just below the national average value of 3.5%.<sup>9</sup> None of the 4 patients who died was identified as having PF (although it is unknown as to whether these patients would have gone on to develop it).

The role of fasciotomy remains controversial. Proponents of fasciotomy argue that it reduces amputation rates.<sup>4,5</sup> None of our patients underwent fasciotomy. Our rate of major limb amputation (33%) compares favourably to published figures (33.3-80%),<sup>1,4,7</sup> supporting our premise that the patchy nature of the disease means that fasciotomies are of limited value.

Other proposed methods for minimizing limb amputations include arteriolytic, hyperbaric oxygen and thrombolytics, some of which are associated with high morbidity, precluding their widespread adoption.<sup>6,8</sup>

Markers of disseminated intravascular coagulation have been shown to be predictors of increased mortality.<sup>10</sup> Our data has shown that patients with a low admission platelet count or increased APTT or INR were more likely to develop PF and require amputations. These findings may be useful indicators for predicting a high risk sub-group of patients, for whom the above therapies could be investigated prospectively.

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