

Introduction

- In 2015, a total of 1,157 cases (43.2%) from 2,677 cases of bleeding disorders were diagnosed as haemophilia A registered in Haemostasis Laboratory in the National Blood Centre Kuala Lumpur (NBCKL).
- Inhibitor formation is one of the primary complications in the treatment of haemophilia A, which causes the bleeding to become more difficult to control and does not respond to the standard therapy
- About 10% of patients with haemophilia A had developed inhibitor in Malaysia.
- Immune tolerance induction (ITI) is the definitive treatment and the only proven therapy to eliminate the inhibitor.
- NBCKL had also offered some of the severe haemophilia A with inhibitor patients with ITI therapy since 2002

Objectives

- ⊗ To describe the proportion of treatment outcomes among patient who had undergone the immune tolerance induction therapy.
- ⊗ To study the factors associated with outcomes in Severe Hemophilia A with an Inhibitor that received immune tolerance induction therapy in National Blood Center, Kuala Lumpur.

Materials and Methods

- A retrospective cross sectional study design was carried out over a period of 16 months from 1st July 2015 till 30th November 2016.
- The study done by reviewing the 18 files of patients diagnosed as haemophilia A with inhibitors underwent ITI therapy at the NBCKL from 2002 till June 2016.
- Data were obtained from patients's clinical notes and recorded into research proforma.

Conclusions

- This study has demonstrated that ITI therapy can be an effective treatment as inhibitor eliminator and can be used as guidance to decide a good selection of patient, the best ITI regime, and the best time to start the ITI therapy for their patient in the future.
- The successful ITI factors includes :
 - Low historical peak inhibitor titre and peak inhibitor titre during ITI (<200BU/ml).
 - Low inhibitor titre before and at start of ITI (<10BU/ml).
 - Start ITI early once inhibitor has been developed. The shorter duration of starting ITI therapy after inhibitor detection could influence the ITI outcome with higher success rate.

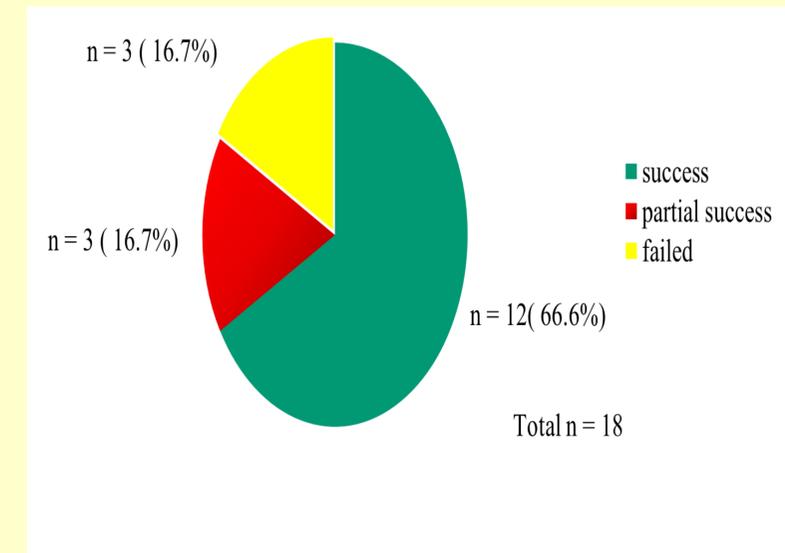
Results

Characteristics of patient's age with the disease and treatment

Variables	n (%)	Median	IQR value
Median age at haemophilia diagnosis (months)	18 (100.0)	9.00	5.25-14.00
Median age at inhibitor detection (years)	18 (100.0)	2.05	1.25-6.03
Median age at start ITI (years)	18 (100.0)	2.45	1.68-11.80

- The youngest patient's age upon diagnoses of at haemophilia A was at 2 months old and the oldest at 52 months old.
- The age at inhibitor detection was as early as 0.7 years old and the oldest was 13.9 years old.
- The age at start of ITI therapy was 1.1 years old as a the youngest age and the oldest age was 15.9 years old.

Number of patients based on ITI outcomes



Factors associated with ITI outcomes

Characteristics	Outcomes			p-value
	Successful	Partial success	Failed	
^b Historical peak inhibitor	10.00 (3.95,22.00)	170.00 (22.00,280.00)	45.00 (35.00,500.00)	0.015*
^b Peak inhibitor ITI	11.00 (2.78,90.00)	200.00 (180.00,220.00)	200.00 (200.00,320.00)	0.018*
^b Inhibitor prior ITI	8.00 (2.60,11.00)	22.00 (20.00,150.00)	10.00 (10.00,100.00)	0.036*
^b Inhibitor start ITI	2.40 (0.00,7.25)	45.00 (12.00,150.00)	10.00 (5.50,200.00)	0.011*
^b Duration between inhibitor detection and ITI (days)	15.00 (0.00,135.25)	210.00 (150.00,4015.00)	480.00 (50.00,3615.00)	0.046*

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