Inhibitor Incidence in A Cohort Of Black South African People With Severe Haemophilia A is Not Higher than that of Caucasian Patients.

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Background

- Inhibitor development is the most serious complication of replacement therapy in haemophilia(1).
- Risk factors for inhibitor development are many and include ethnicity(2).
- Some studies suggests that people of African descent may have a higher risk of inhibitor development compared to Caucasians(3).
- The South African haemophilia population consists of 80% Black Africans and data on previously untreated patients (PUPs) have never been published.

Objectives

To document the incidence of inhibitors in PUPs with severe haemophilia A from a large comprehensive care center in South Africa.

Patients and Methods

❑ **Study design**: This study was a single centre retrospective cohort study

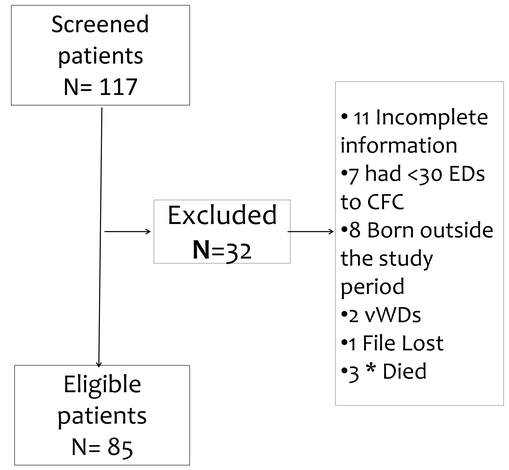
❑ **Study population**: comprised of patients that met the following inclusion criteria:

- PUPs with severe haemophilia A born; between 1989-2010; must have complete clinical records especially for first exposure day and peak ; treatment at first exposure day; follow up must be up to 50 exposure days(EDs).

❑ **Procedures**

- Demographic information, family history of inhibitors, inhibitor test results, inhibitor risk factors , genetic analyses were extracted from the hospital patient records, clinical laboratory information system and genetic counselling files.
- Data was anonymized and coded prior to analysis.
- Data was analyzed descriptively

Figure 1 Patient flow chart



EDs= Exposure days, CFC = Clotting Factor Concentrate, vWD = Von Willebrand's Disease * The three patients who died, did so before reaching 30 EDs

Table 1. Population parameters(n=85)

Parameters	Inhibitor Patients(n=15)	Non-inhibitor patients (n=70)
Ethnicity, n		
- Blacks	12	43
- Whites	2	24
- Others	1	3
Family history of haemophilia, n	9	42
Family history of inhibitors, n	1	1
Type of CFC (pdCFC/rCFC)	All pdCFC	All used pdCFC
Intensity of treatment, n	0	13
Mean Exposure days, d	114	142

Results

- 85 patients met the eligibility criteria. **See figure 1**
- A clinically significant inhibitor was noted in 17,6% which comprised 15/85 of the study cohort. **See table 1**
- 80% were low titre and 20% were high titre inhibitors.
- 80% were black, 13% white and 7% other ethnicities.
- The mean number of exposure days was 114.
- These participants were on demand CFC replacement.
- There was no coincidence of inhibitor development with viral infections, and periods of surgical interventions.
- The inhibitors persisted during the follow up visits and required bypassing agent treatment or immune tolerance induction

Discussion

- In a number of published studies, non-Caucasian participants who develop inhibitors comprised of Latinos, Hispanics and African American populations but no black Africans

- In this cohort, black Africans comprised the majority (67%, n=57/85) of participants. Of the patients that developed inhibitors, 80%(n=12/15) were Black.

Conclusion

The inhibitor incidence of 17.6% in our mainly black African patients was similar to that seen in Caucasians in published studies and not higher as seen in African Americans and Latinos in published studies. Further studies are required to confirm this retrospective finding.

References

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Disclosures

Authors have no conflict of interest to declare