Stroke prevalence amongst sickle cell disease patients in Nigeria: a multi-centre study

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Abstract:

Background: Stroke is a life-changing, debilitating complication of sickle cell disease (SCD). Previous studies had recorded high stroke prevalence amongst this group of patients. Nigeria has a large population of people affected by this condition and this study aims to assess the stroke prevalence in this large population.

Methodology: Stroke prevalence data from 14 physicians working in 11 tertiary health centres across the country was collated by doctors using the sickle cell registers and patient case notes. This data was then collated and used to obtain the overall stroke prevalence in adult and children.

Results: The stroke prevalence in sickle cell disease patients in Nigeria was observed to be 12.4 per 1000 patients. Prevalence in the adult patients was 17.7 per 1000 patients and 7.4 per 1000 patients in children. Twenty three percent of the affected patients had more than stroke episode.

Conclusion: The stroke prevalence in Nigeria is lower than previously recorded rates and further studies will be required to investigate other factors which may play a role.

Keywords: sickle cell, stroke, Nigeria, prevalence

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Introduction:

Stroke is a well-known debilitating complication of sickle cell disease. It is a leading cause of death in children as well as adults with this condition, with a reported risk of first cerebrovascular accident (CVA) in the first 20 years of life being 0.761 per hundred patient years[1]. A Jamaican cohort study reported a prevalence rate of 7.8% in 310 patients of all genotypes aged

between 9 and 17 years[2]. Other cohort studies revealed an incidence of first stroke of 500 - 1,000/ 100,000 person-years[3,4]. The relatively high prevalence rate, of this catastrophic complication in this young group of patients, has been the reason behind the adoption of screening programs, conducted early in life to detect and prevent stroke in sickle cell disease (SCD).

Nigeria has a population of 160 million, and an estimated sickle gene carrier rate of about 40%, with about 150,000 children being born annually with sickle cell disease[4], and should be at the fore front of such preventive programs. There are various environmental, as well as genetic risk factors, which may protect or predispose patients to the development of stroke.

Stroke in sickle cell is a preventable cause of morbidity and mortality in this group of patients. Affected persons are in most cases unable to carry on with their day to day activities. This leads to permanent disability in which case one or both Parents have to be tied down with care for the patient. Lifelong disability in children especially in the African setting leads to worsening poverty, poor quality of life, reduced standard of care and higher probability of recurrence.

Routine Trans-cranial Doppler ultrasound screening in patients with SCD is currently not done in any part of the country. Therefore, the country needs a national policy on prevention of stroke secondary to SCD, and data collection to determine its prevalence is a prerequisite. The objective of this study was to evaluate the prevalence of stroke among SCD patients attending health institutions in Nigeria.

Patients and Methods:

Nigeria is a West African country, bordered by Togo, Republic of Benin, Cameroon, Chad and the Atlantic Ocean. It is divided into 6 geo-political zones which have 36 states. It is the most populous African nation with an estimated population of 150 million.

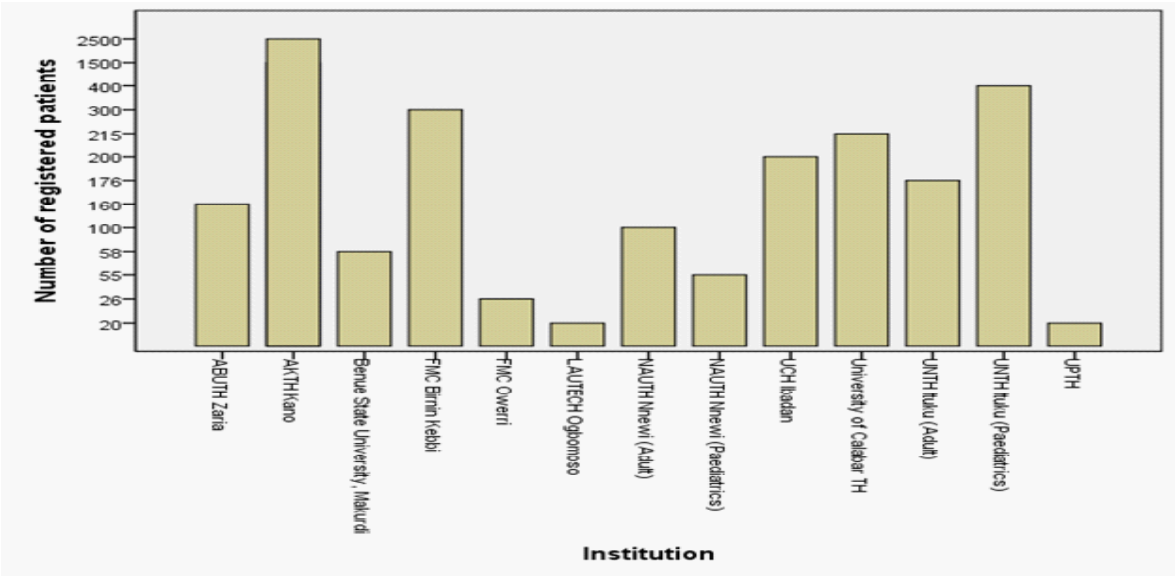
An 8-item pre-validated questionnaire was sent to 22 Physicians (Haematologists and Paediatricians) in 17 tertiary health institutions across the different geo-political zones in Nigeria. Selection of the participating centres was non-randomized and questionnaires were sent to all centres which the authors had correspondence with. Information included; specialty, number of registered patients, number of patients diagnosed with stroke, and number of patients with recurrence. Other information obtained included the types of stroke and mean age of the affected patients as well as whether the physician feels there was a need to screen all SCD patients using TCD.

Information was obtained from 14 physicians working in adult or paediatric sickle cell units in 11 tertiary health institutions across the country. Some of the participating centres had separate adult and children clinics, while some had a joint sickle cell clinic. These physicians obtained some of the data from the sickle cell registers and case notes in each centre. This data was used to estimate the prevalence of stroke in Nigeria. Data obtained was collated and expressed in tables and figures, prevalence and incidence rates were determined using IBM SPSS 19.0 (United Kingdom 2011).

Results:

A total of 5,721 sickle cell patients were registered in 14 sickle clinics in 11 tertiary health institutions across the country. This ranged from 20 to 2500 patients with a mean of 409 and a median value of 168 patients per

Figure 1. Distribution of patients recorded in tertiary health institutions across Nigeria

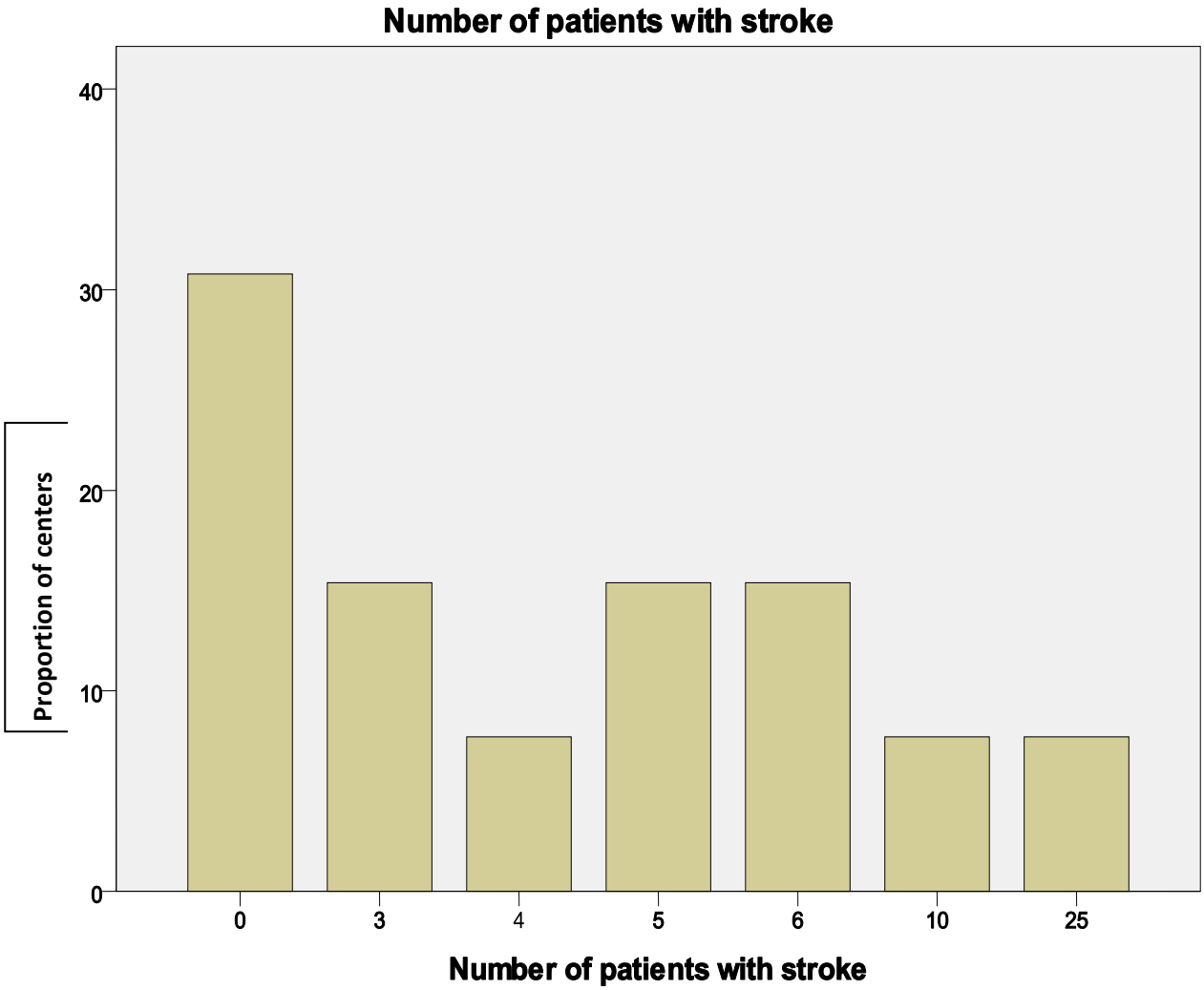


centre. Figure 1 contains the number of registered sickle cell patients recorded in each of the participating centres.

The duration of experience of the Physicians working with sickle cell patients and length of time over which this data was captured ranged from 6 to 16 years, with a mean duration of 11.1 years(SE of mean 1.4) and median of 10 years.

There was no stroke observed in 3(21.4%) of the 14 centres, while 1(7.1%) and 2(14.3%) centres each had 5

Figure 2. Frequency distribution of number of patients who had stroke across the participating centers

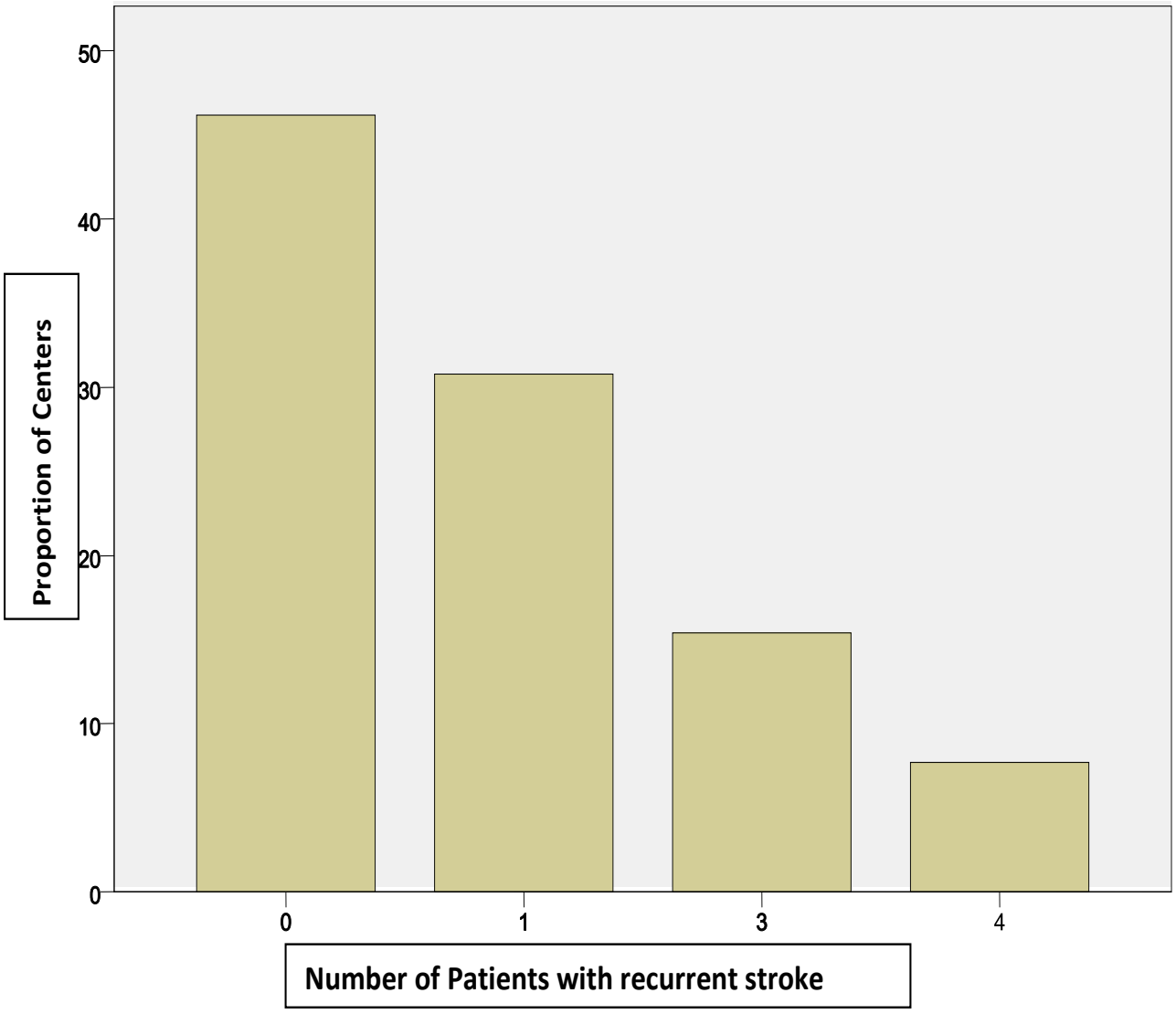


and 6 patients with stroke respectively. Only one centre recorded as high as 25 patients with stroke – see Figure 2.

Some (there are 6 out of 14 centres) centres (42.9%) 6/14 did not record/document multiple strokes in their registered patients, while 4/14 (28.6%) of the centres recorded only 1 patient with more than one episode of stroke – see Figure 3.

The prevalence rate of stroke in sickle cell was determined to be 12.4 per 1000 sickle cell patients, obtained from the 14 centres who participated in the study. Of the 71 patients who had stroke, 17 had had more than one episode of stroke, giving a 23.9% recurrence rate and incidence of multiple strokes of 2.97 per 1000 patients. In the study, there were 2955 paediatric patients studied

Figure 3. Frequency distribution of the number of patients who had more than one stroke in Tertiary health institutions recorded



across 3 centres, with a prevalence rate of 7.4 per 1000 patients and 2766 adult patients with a prevalence of 17.7 per 1000.

The average age incidence of stroke across all the centres was 21 years in adults and 6 years in the children observed in the study. The prevalence of stroke varied from zero in the adults seen at Nnewi and Port Harcourt

Table 1. Stroke prevalence in patients of each Health Institution

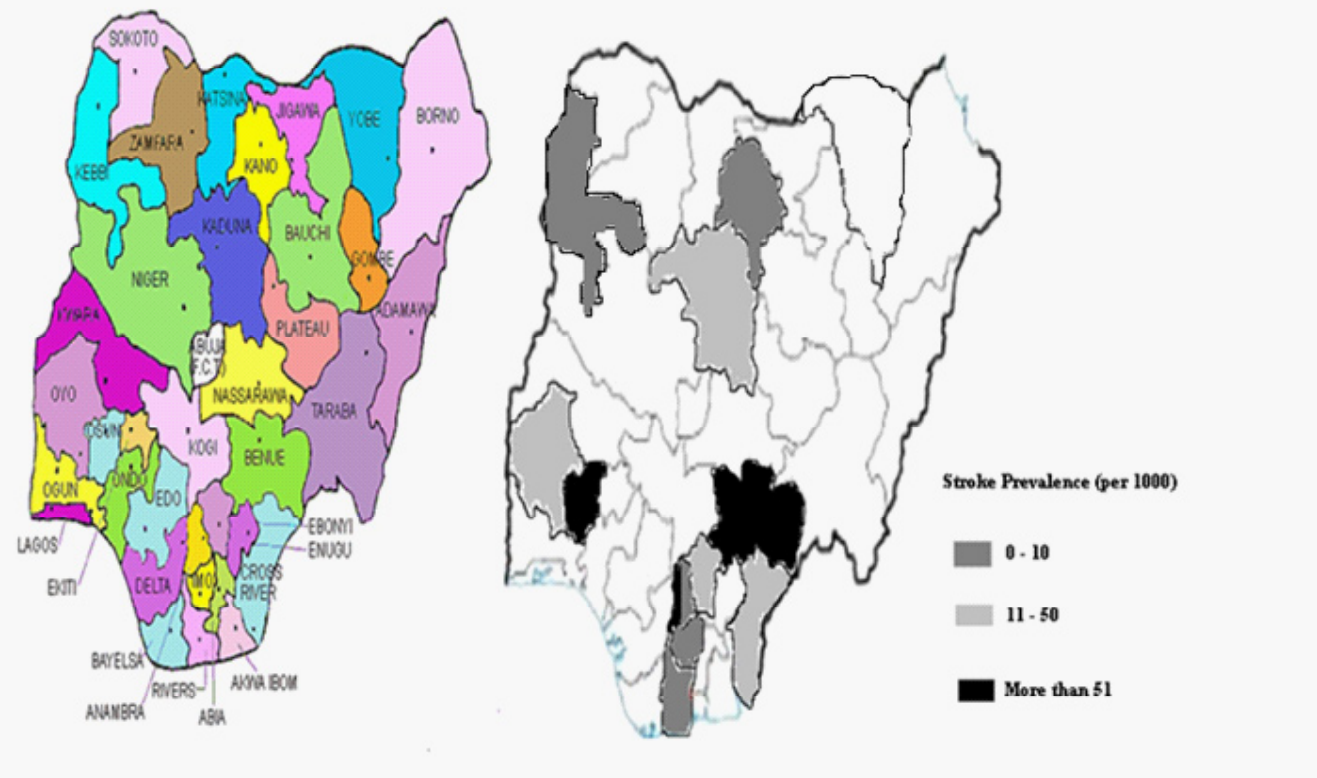
Institution (Geo-political zone)	No of registered patients	Stroke prevalence (per 1000 sickle cell patients)
Ladoke Akintola University Teaching Hospital, Ogbomoso (Adult) - (South West)	20	150
Benue State University Teaching Hospital, Makurdi (Adult) – (North Central)	58	116
Ahmadu Bello Teaching Hospital Zaria (Adult) – (North East)	160	25
University of Portharcourt Teaching Hospital (Adult) - (South South)	20	0
University of Nigeria Teaching Hospital Enugu State (Paediatrics) – (South East)	400	22.5
University of Calabar Teaching Hospital (Adult) - (South South)	215	27.9
Federal Medical Center Owerri (Joint) - (South East)	26	0
University of Nigeria Teaching Hospital Enugu State (Adult) – (South East)	178	33.7
Nnamdi Azikiwe University Teaching Hospital Nnewi (Paediatrics)- (South East)	55	54.4
Nnamdi Azikiwe University Teaching Hospital Nnewi (Adult)- (South East)	100	0
Aminu Kano Teaching Hospital Kano (Adult) –(North West)	1500	6.7
Aminu Kano Teaching Hospital Kano (Paediatrics) – (North West)	2500	1
University College Hospital Ibadan (Adult) - (South West)	200	25
Federal Medical Center, Birnin Kebbi (Joint clinic) – (North West)	300	6.6

to as high as 150 per 1000 patients in Ogbomoso, Osun State – Table 1.

See figure 4 for the stroke prevalence map of the country.

With regards to trans-cranial Doppler (TCD) screening for stroke 92.9% (13/14) of the responding physicians

Figure 4. Prevalence Map of stroke in patients with SCD in Nigeria



felt it was important to implement this as a preventive measure. Only 1 haematologist felt it was not necessary to screen all sickle patients with TCD.

Discussion

Sickle cell disease, an inherited haemoglobin disorder leads to chronic multi-organ complications. The frequency of these complications increases with age[5]. With improved health care and the advent of neonatal screening, the prevalence of such chronic debilitating complications such as stroke needs to be evaluated in the teeming Nigerian population. Previous studies done in Nigeria were done in a single centre, but a multi-centre study would definitely give a more robust view of the problem. The overall prevalence of stroke in this study was found to be 12.4 per 1000; this is lower than the value obtained by George *et al* in Portharcourt[6]. Other studies done in Jamaica by Balkaran *et al*, with 310 patients showed a prevalence of 7.8%[2] and 4.01% by Ohene-Frempong *et al*[3]. Frempongs study was a longitudinal study carried out in sickle cell patients in America, whose phenotypic expression of this complication may be exacerbated by several dietary as well as environmental factors.

However the prevalence amongst paediatric patients obtained in this study were 7.4 per 1000, this is similar to the prevalence rates obtained by Izuora *et al* [7]in Enugu, where a rate of 6.5 per 1000 was determined. Studies done by Lagunju *et al* in the University college hospital Ibadan, South Western Nigeria, revealed 8.4% (84 per 1000) of the paediatric patients had stroke, though a higher percentage had neurological complications[8]. This may also be the effect of environmental factors as well as other genetic factors unique to each tribe or population, which may be at play in the occurrence of stroke. This study revealed higher stroke prevalence amongst the adult population compared with children and; this is contrary to previous data. However, in as much as this

may be a different trend it can also be explained by the high mortality rate in children in the country which prevents those that have the predisposition to stroke to grow to adulthood.

Majority of the patients observed in this study were from the Northern part of the country and this reflects the distribution in prevalence of sickle cell across the country, which shows wide variability. Previous studies in Nigeria had pointed out this variability in prevalence as well as in the type of haemoglobin abnormalities across the country, with a high frequency of haemoglobin C occurring in the South-western part of the country and to a lesser extent in the Southeast[8].

The recurrence rate for stroke in this study is much lower than had been recorded in previous studies, with values as higher as 67%[1] and 90%[9]. Worthy of note is that these studies were carried out before the advent of transfusion programs as a preventive modality in stroke patients. Some of the patients in the study population who had had stroke were already on a preventive transfusion program, and this explains the lower recurrence rate noted, though some of them with limited access to healthcare and safe blood, were not.

Accounts of several studies around the globe show markedly high prevalence of stroke in sickle cell patients of up to 4 - 5%[10]. This study has shown that Nigerian patients have a much lower prevalence. This has been supported by the clinical experience of the authors and suggests that other genetic or environmental factors may be at play and require to be further investigated.

Conclusion

The stroke prevalence amongst sickle cell disease patients in Nigeria was observed to be 12.4 per 1000 patients. Prevalence in the adult patients was 17.7, and 7.4 per 1000 patients in children. The prevalence of stroke in Nigerian sickle cell patients is lower than had been observed in other stroke studies. Further studies will be required to investigate the genetic as well as environmental factors which may be responsible for this disparity.

Limitations of the study:

Low response to sent questionnaires was a major setback in this study.

Disclosures:

None

Conflict of interest:

There was no conflict of interest amongst the authors of this manuscript

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