



Keratocystic odontogenic tumours of the jaws and associated pathologies: A 10-year clinicopathologic audit in a referral teaching hospital in Kenya[☆]

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ABSTRACT

Aim: To establish the pattern of occurrence and the clinicopathological features of keratocystic odontogenic tumour (KCOT) over a 10-year period.

Materials and method: Patients from the University of Nairobi Dental Hospital treated for KCOT were included in the study over a 10-year period. The study highlights the demographic, clinico-radiological and histological features of these tumours.

Results: A total of 22 confirmed cases of KCOTs were recorded with equal gender prevalence; (M:F = 1.44:1). The age range of the patients was from 10 to 69 years with a peak in the second decade of life (mean = 27.5 yrs). Of the 22 cases, 15 (68.2%) occurred in the mandible of which eight (53.3%) involved the body, five (33.4%) the angle and ramus. Six (27.3%) occurred in the maxilla, and one (4.5%) was in both jaws and was associated with Gorlin–Goltz Syndrome. The most common presenting complaint in most patients was swelling 54.6%, and in 18.2% was incidental finding. Eight (36.4%) cases showed satellite cysts upon pathologic evaluation. Thirteen (59.1%) cases were managed by surgical excision, while nine (40.9%) were managed by enucleation.

Conclusion: Based on the outcome of this study, KCOT present mostly in body, angle and ramus of the mandible and its peak is in the second decade of life.

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1. Introduction

Keratocystic odontogenic tumour (KCOT), formerly known as odontogenic keratocyst (OKC) is a benign unicystic or multicystic intraosseous neoplasm of odontogenic origin, which arises from remnants of dental lamina or glands of Serres (Browne, 1971; Kolokythas et al., 2007; Gonzalez et al., 2008). It has a potential for locally destructive behaviour, recurrence, and a tendency for multiplicity particularly when associated with nevoid basal cell carcinoma syndrome (NBCCS) (Cawson et al., 1998; Slootweg, 2007). It has a peak incidence in the second and third decades of life and a gradual decline thereafter with a male dominance and has a considerable predilection for the posterior body of the mandible and ascending ramus (Brannon, 1976; Ahfors et al., 1984; Myoung et al., 2001; EL-Gehani et al., 2009). Patients present with

swelling, infection, pain, paraesthesia, cellulitis, and trismus (Chirapathomskul et al., 2006). It has exceedingly high recurrence rate due to the delicate thin epithelial lining, the presence of satellite/daughter cysts in the wall, anteroposterior growth in cancellous bone with finger like projections and a high mitotic potential (Maurette et al., 2006). Radiographically it is a unilocular or multilocular well circumscribed radiolucent lesion with scalloped and corticated margins (Browne, 1971; Poramate et al., 2010). Histologically, the tumour has an epithelial lining that is uniformly thin, generally ranging from 8 to 10 cells layers thick. However, they lack rete ridges and often have an artifactual separation from their basement membrane. The basal layer has palisading cells with polarized and deeply staining nuclei of uniform diameter. The wall is fibrous and contains satellite cysts which can grow and form individual cysts. The luminal epithelial cells are parakeratinized and produce an even corrugated profile. Focal zones of orthokeratin have been described. Also, epithelial proliferations similar to ameloblastoma have been reported (Crowley et al., 1992; Slootweg, 2007; Poramate et al., 2010).

This paper highlights the pattern of occurrence and the clinicopathological features of KCOT in patients who were treated at the

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University of Nairobi Dental Hospital (UONDH), over a period of 10 years. UONDH is a centre of excellence of training of oral and maxillofacial surgeons.

2. Materials and method

The data for this analysis was obtained from the records of all the cases of KCOTs treated at the UONDH from 1999 to 2009 (10 years). The majority of such cases present to the department of Oral and Maxillofacial surgery, oral pathology and oral medicine for management from all over the country. The inclusion criteria were patients diagnosed with KCOT at the UONDH. Patients with incomplete records and those with disputed histopathology findings were altogether excluded from the study. The records were examined and the following information recorded; demographic characteristics, age, gender, site of lesion, radiographic and histopathological information. Archival histological blocks and slides of the cases were retrieved and the primary diagnosis was approved by re-evaluation of the biopsies by two experienced Oral Pathologists. The pathologists concurred on 22 out of 61 cases initially diagnosed as KCOT. The rejected cases were mainly unicystic ameloblastomas.

Radiographic findings were recorded from orthopantomograms (OPGs) and Computed Tomographic (CT)-scans. Treatment consisted of marginal resection (and disarticulation where indicated) with subsequent reconstruction with bone graft secured with stainless steel or titanium reconstruction plates.

3. Results

During the 10-year period, a total of 22 tumours were recorded as KCOT. These were re-evaluated by an independent external oral pathologist, Prof. Slootweg (the seventh author) and confirmed to be KCOT. The study population comprised 13 (59.1%) men and nine (40.9%) women (female to male ratio of 1–1.44). The age range was from 10 to 69 years (mean = 27.5 yrs). KCOT had a peak occurrence in the second (31.8%) and third (27.2%) decade of life (Table 1).

Fifteen (68.2%) lesions occurred in the mandible, out of which two (13.3%) were in the anterior region while 13 (86.7%) occurred in the posterior region. Eight (53.3%) were present in the body and five (33.4%) at the angle and ramus region (Fig. 1). Six (27.3%) occurred in the maxilla; five (83.3%) presented in the anterior region and one (16.7%) in the premolar region. In one case, the lesion had developed in the maxillary sinus. The remaining KCOT (4.5%) occurred both in the mandible and in the maxilla and was associated with Gorlin–Goltz Syndrome. It occurred in a 20-year-old male at the time of initial diagnosis. The patient also presented with multiple basal cell carcinomas of the skin, bifid ribs and had intracranial calcifications upon radiographic evaluation. The follow-up period for this patient was 36 months.

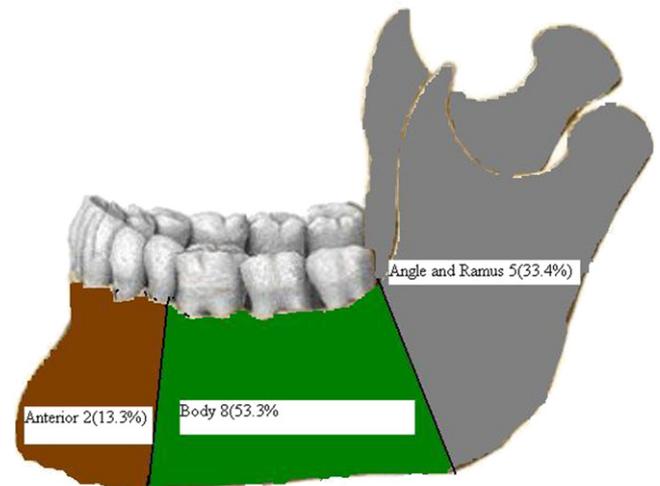


Fig. 1. Schematic representation of the lesions involving the mandible. All the lesions were unilateral. (Note: demarcated areas represent anatomical regions involved).

The majority of the patients 18 (81.8%) complained of swelling, pain, discharge, loosening and displaced teeth. Only four (18.2%) were discovered incidentally during routine dental examination. The most common complaint of patients at the time of admission was swelling 12 (54.6%) while pain was reported in 3 (13.6%) lesions (Table 2).

In 2 (9.1%) cases of KCOT, the lesions were associated with an impacted tooth; a mandibular third molar and a maxillary canine (Fig. 2A and B). Multilocular radiolucency (Fig. 3A and B) was the most common radiographic finding 13 (59.1%) while, unilocular radiolucency occurred in 9 (40.9%) of the patients.

Of the KCOTs studied, 18 (81.8%) showed pure parakeratinization (Fig. 4A and B) and four (18.2%) had both mixed parakeratinization and orthokeratinization. There was palisading of the basal layer in all the tumours (Fig. 4B). Parakeratinization was present in those that showed multiple lesions. There were 36.4% with satellite cysts and 4.5% showed epithelial budding (Fig. 5A and B).

Only one (4.5%) patient, a 30-year-old male presented with recurrence 24 months after enucleation. The tumour was noted to be parakeratinized and had one or more satellite cysts with features of epithelial budding.

An audit of the treatment used for the lesions showed that 13 (59.1%) were managed by surgical resection and nine (40.9%) by enucleation followed by reconstruction using titanium or stainless steel plates (Fig. 3C).

4. Discussion

There is no information regarding patient details of KCOT in a Kenyan population. A male predominance has been reported for KCOT in previous studies, the general distribution being 60% male and 40% female. The sex distribution in this study was similar to that by Myoung et al. (2001); we confirmed a male predominance

Table 1
Age and gender distribution in patients with KCOT.

Age in years	Male (%)	Female (%)	Total (%)
1–9	0 (0.0)	0 (0.0)	0 (0.0)
10–19	4 (18.2)	3 (13.6)	7 (31.8)
20–29	4 (18.2)	2 (9.1)	6 (27.2)
30–39	2 (9.1)	3 (13.6)	5 (22.7)
40–49	2 (9.1)	0 (0.0)	2 (9.1)
50–59	0 (0.0)	0 (0.0)	0 (0.0)
60–69	1 (4.5)	1 (4.5)	2 (9.0)
70–79	0 (0.0)	0 (0.0)	0 (0.0)
80–89	0 (0.0)	0 (0.0)	0 (0.0)
90–99	0 (0.0)	0 (0.0)	0 (0.0)
Total	13 (59.1)	9 (40.9)	22 (100)

Table 2
Presenting symptoms of KCOTs.

Symptoms	Frequency	Percentage (%)
Swelling	12	54.6
Swelling + loose + displaced and loose teeth	3	13.6
Swelling + pain	3	13.6
Incidental diagnosis	4	18.2
Total	22	100

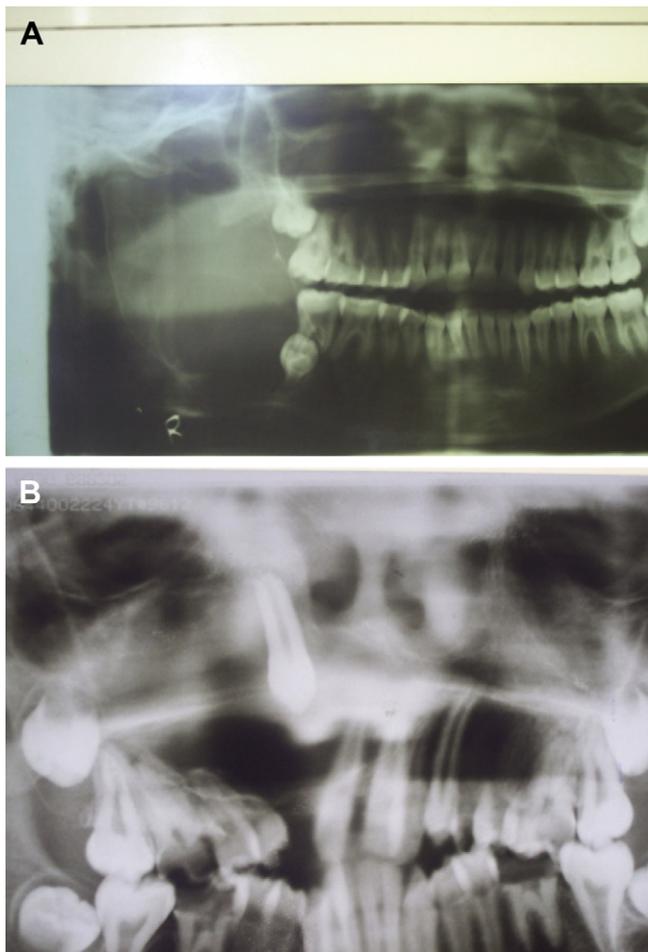


Fig. 2. Panoramic radiography demonstrates a unilocular radiolucency associated with an impacted third molar in the fourth quadrant (A) and an impacted canine in the right maxillary sinus (B).

(Myoung et al., 2001). Although the mean age in this series corresponded to those in other studies, it was relatively lower (Brannon, 1976; Ahfors et al., 1984; Myoung et al., 2001; Morgan et al., 2005; Maurette et al., 2006; Poramate et al., 2010). Unlike other studies in this field, which mostly reported the third decade of life as the most common age of occurrence, most of the patients in this study were young with a peak occurrence in the second decade of life (Myoung et al., 2001; Chirapathomskul et al., 2006; Maurette et al., 2006; EL-Gehani et al., 2009; Poramate et al., 2010). This was different in this study because patients, in our set-up, tend to seek treatment when symptoms are advanced.

The mandible was involved more frequently than the maxilla, which is comparable to the figures reported by others, ranging from 65% to 83% for the mandible, (Brannon, 1976; Philipsten et al., 2005; EL-Gehani et al., 2009; Poramate et al., 2010). The findings of this study compared favourably with other studies, which reported the mandibular body and post-molar region (angle and ramus) as the most common site of occurrence (Ahfors et al., 1984; Crowley et al., 1992; Myoung et al., 2001; Chirapathomskul et al., 2006; EL-Gehani et al., 2009; Poramate et al., 2010). With regard to symptoms, several researchers have reported that 50–90% of KCOTs are symptomatic at the time of diagnosis, which is comparable to our finding of 81.8% (Brannon, 1976; Crowley et al., 1992; Morgan et al., 2005; Chirapathomskul et al., 2006).

Swelling, combined with pain, pus discharge, or both were the chief presenting symptoms in line with other investigators

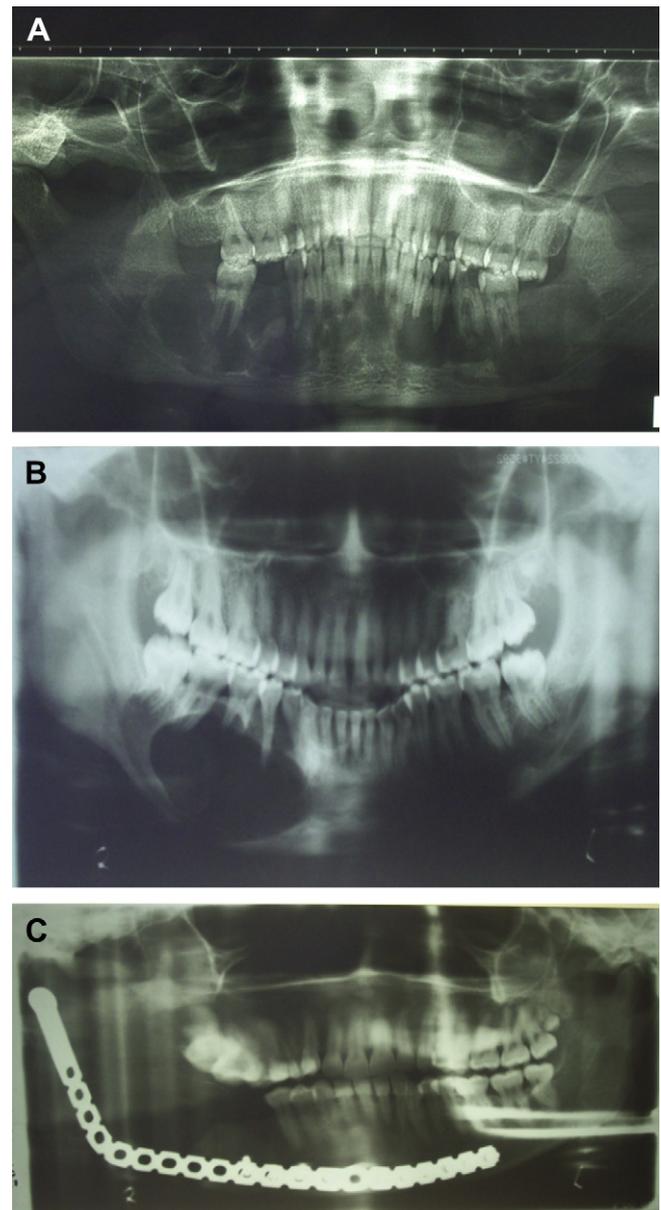


Fig. 3. Panoramic view demonstrates multilocular radiolucency of the mandible (A & B). Reconstruction of the mandible of a patient who had undergone surgical resection of an extensive KCOT (C).

(Brannon, 1976; Crowley et al., 1992; Myoung et al., 2001; Poramate et al., 2010). The association of KCOT with unerupted teeth in this series was lower than reported (Neville et al., 2002; EL-Gehani et al., 2009). This is because some of the radiographs of patients referred from other hospitals could not be retrieved.

A characteristic regular parakeratinized stratified squamous epithelium and well-defined basal layer are among the important histological features distinguishing KCOT from other jaw cysts (Philipsten et al., 2005). Also, epithelial proliferations similar to ameloblastoma have been reported (Slootweg, 2007).

Parakeratinizing lesions have accounted for the majority of KCOTs in series conducted in different countries, such as USA and France (Crowley et al., 1992; Poramate et al., 2010). In this study, 81.8% of KCOTs were parakeratinized which is compared to that of Gonzalez et al. (2008) (94%). The presence of one or more daughter cysts adjacent to the cystic wall of the tumour demonstrated in this

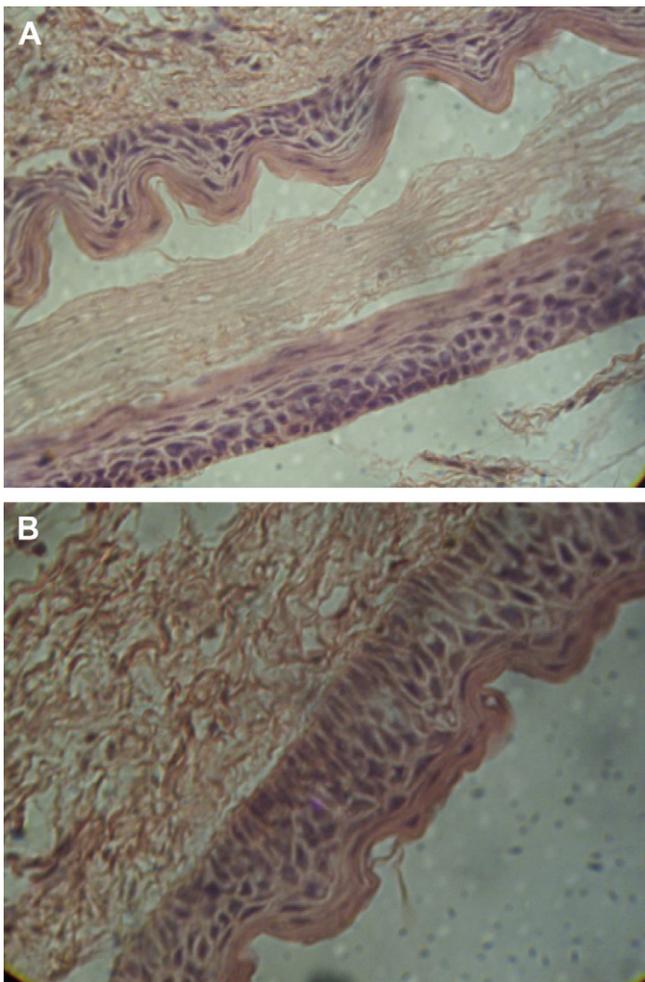


Fig. 4. Prominently palisaded basal cell layer with dark-staining nuclei and a corrugated surface with parakeratinization undergoing desquamation into the cyst lumen (A) (H-E staining, original magnification 4 \times), same lesion (B) (H-E staining, original magnification 10 \times).

series (36.4%) is considerably closer to the figure reported by Myoung et al. (2001) (30%). The above criteria were used in the histopathological review of this series of KCOT cases, and cystic lesions which did not consistently show these features were excluded from the study.

The recurrence rate in this series is within the range noted from the studies, 2.5% to as high as 62.5% (Forssell et al., 1988; Gonzalez et al., 2008; Poramate et al., 2010). These findings were consistent with those of Forssell et al. (1988) and Habibi et al. (2007) who reported a higher recurrence rate in children and young adult patients. This may be attributed to the fact that younger patients usually receive more conservative treatment, which may lead to higher recurrence. Myoung et al. (2001) have documented that the site of involvement significantly affects the recurrence rate. Although the current study did not find any significant relationship between lesion location and recurrence, the recurrence commonly occurred in the mandibular molar and post-molar region.

A diagnosis of association of Gorlin–Goltz with KCOT in this series was similar to those in other reports, which range between 1.4% and 8.2% (Gorlin et al., 1965; Gustafson et al., 1989; Crowley et al., 1992; Gonzalez et al., 2008). Other findings report that KCOTs associated with NBCCS have a higher recurrence rate (Habibi et al., 2007). The results of this series did not confirm such an association as the recurrent lesion was not associated with NBCCS.

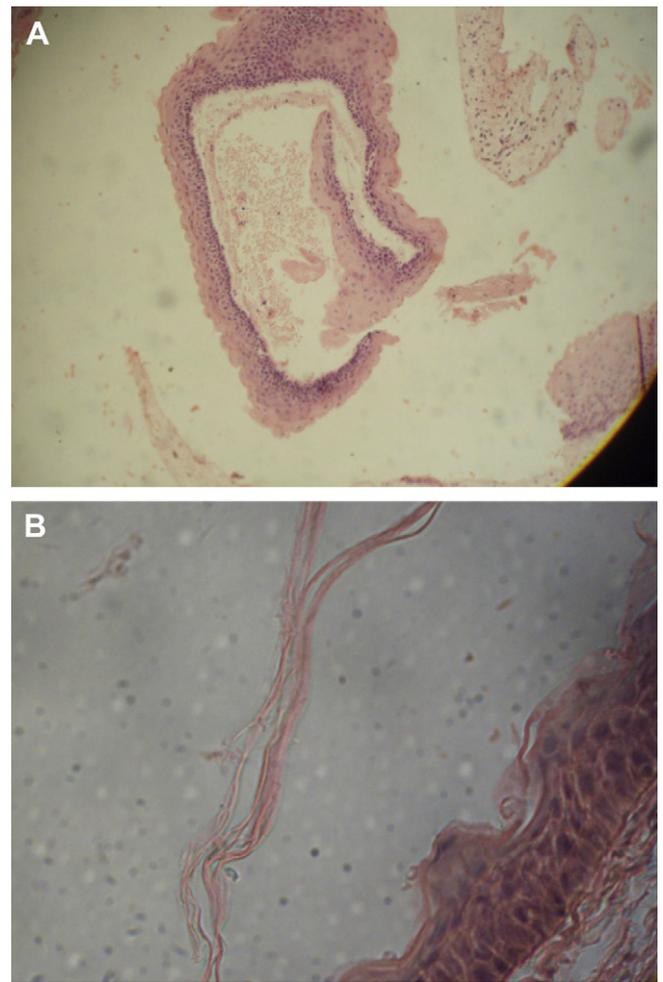


Fig. 5. Prominent daughter cysts containing keratin whorls (A) (H-E staining, original magnification 4 \times), budding of tumour epithelium with koilocytosis (B) (H-E staining, original magnification 10 \times).

However, since the latter patients have a considerable tendency to grow new lesions, a longer follow-up may be necessary (Morgan et al., 2005).

The treatment of choice for KCOT lesions is still debatable. Traditionally, enucleation followed by peripheral localized osteotomy is considered the best treatment for KCOTs (Chirapathomskul et al., 2006; Maurette et al., 2006). However, its high surgical morbidity and relatively high rate of associated recurrence mean that it cannot be considered the most ideal form of surgical management (Morgan et al., 2005; Chirapathomskul et al., 2006). Recent studies have revealed that marsupialization is applicable as a conservative technique for large lesions (Maurette et al., 2006; Maker et al., 1996) while Poramate et al. (2010) suggest that enucleation with the aid of CT and adequate postoperative vigilance is a conservative treatment which yields clinically acceptable results.

In this series, only one enucleated lesion showed recurrence, which is somewhat closer to those in other studies (Myoung et al., 2001; Morgan et al., 2005; Chirapathomskul et al., 2006). Since in this study the mean follow-up period was relatively short compared to other studies, the number of recurrent lesions was relatively small. Therefore, long follow-up periods are suggested for this tumour and as research continues the treatment for KCOT may involve molecular-based modalities which will reduce or eliminate the need for aggressive surgical management.

5. Conclusion

Based on the findings in this series, the occurrence of KCOT can mimic a cyst or ameloblastoma; clinicians therefore should be vigilant while making a definitive diagnosis.

Recommendation

Multi-centre prospective studies should be conducted in this region to ascertain the occurrence, recurrence and associated pathologies of KCOTs including Gorlin–Goltz Syndrome.

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Conflict of interest

The authors had full freedom of investigation and there were no potential conflicts of interest.

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