A REVIEW OF
THE MANAGEMENT OF CAROTID BODY TUMOURS AT KENYATTA NATIONAL HOSPITAL (JAN 1990 - DEC 2000)

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DECLARATION

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This dissertation is my original work and has not been presented for a degree in any other University.

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DEDICATION

I dedicate this work to my daughter Nereah Atieno Muleshe (deceased), who was elevated to glory in July 1999 as I toiled away from home, laying the foundation of my eventual life-long career -Surgery. May her spirit rest in peace.

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EXECUTIVE SUMMARY

This was a retrospective study covering the period from January 1990 to December 2000 inclusive. It involved a review of hospital records of patients who presented at Kenyatta National Hospital (KNH) during this period and were diagnosed to have carotid body tumour (CBT). The relevant literature on the subject was reviewed and justification given for the study. The epidemiological trend, the pattern of presentation, the diagnostic work-up and the modes of treatment of CBT were covered. The study methodology was outlined, and the data obtained processed and analysed. The results were then presented and discussed, and from them conclusions and recommendations made. It is expected that it will be possible to draw a protocol for the standardized management of these patients at the KNH. The whole study should also form a basis for future works on the subject.

During the eleven year period a total of 30 patients were treated, out of whom 21(70%) were female and 9(30%) male, giving a female to male (F: M) ratio of 2.33:1 or 7:3 A third of the patients came from Central Province and only one (3.3%) from Coast. No patient was seen from North Eastern Province. The youngest patient was 20 years old and the oldest 96 years old. The annual prevalence of the disease was 2.72.Patients, even though the most number of patients (7 ie 23.3 %) presented in 1996. There were no obviously identifiable predisposing factors. All the patients presented with a lump in the neck of variable duration; the most favoured investigation was carotid angiography. Twenty eight
(28) out of the 30 patients (93.3%) were operated on, out of whom only one (3.3%) died during reoperation.

1. INTRODUCTION

Carotid body tumour (CBT) is a rare neoplasm of the carotid body, a mass of chemoreceptive tissue found on both sides of the neck at the bifurcation of the common carotid artery. These tumours are of particular interest, not only because of their rarity but also because of the diverse opinions surrounding their biological behaviour, diagnosis and management [1] The commonest mode of presentation is a longstanding painless, pulsatile, slow-growing mass in the neck region at the angle of the mandible. These characteristics make the correct diagnosis of CBT a major clinical challenge, especially to the lower cadre health personnel i.e. nurses and clinical officers, and general practitioners, who most often are the first to encounter these patients. Such persons may lack the very high index of suspicion, which is paramount as an initial step in the diagnosis of CBT. As such mismanagement of these patients in peripheral hospitals where most are first seen is not uncommon; this may take the form of inadvertent incision or excision of the mass under the erroneous impression that it is an enlarged lymph node, a lipoma or an otherwise benign and innocent growth in the neck.

Obviously such a manoeuvre is catastrophic, often leading to torrential haemorrhage and hence greatly endangering the patient's life. That such patients rarely reach competent hands in the referral hospitals further compounds their fate. The insidious onset of CBT as outlined above makes the patient to seek medical attention late. The onus, therefore, lies with the medical person who encounters such a patient first. A high index of suspicion would help to appropriately and promptly refer the patient accordingly; however the most important step in the proper diagnosis of this condition is a proper clinical examination with the simple but paramount basics of inspection and palpation of suspicious neck lumps. Most of these patients end up at the Kenyatta National Hospital, the only public hospital with the necessary facilities and skilled manpower for managing such patients. This study is expected to create the necessary awareness and guidance for the proper care of patients with CBT.

2. LITERATURE REVIEW

1:1 BACKGROUND INFORMATION

Carotid body tumour (CBT) was first described by von Haller in 1743 [2] . In 1880 Reigner performed the first excision of a CBT [3]; the patient did not survive. In 1886 Maydl [4] removed a CBT; his patient survived but had hemiplegia and aphasia. Albert [5] was the first to successfully excise a CBT without ligating the carotid vessels. In 1903 Scudder [4] reported the first successful removal of a CBT in the United States. There is no published report about the first excision of a CBT in Kenya.

Evidence presented by Arias-Stella [6] indicated that carotid bodies were larger in people living at high altitude (7,000-14,000 ft) in the Peruvian Andes than for those living at sea level. In 1973 Saldanha and associates [7] reported that in a Peruvian population CBTs were ten times more frequent among patients from high altitudes than among those living at sea level. Controversy exists over the incidence of malignancy in CBT, with reports in the literature varying from 7 to 100 percent [8]. Metastases should not be confused with paragangliomas that are multicentric in origin [9].

Romanski [10] stated that histological appearance of the tumour is not a guide to the ultimate course it might pursue. At the moment these tumours are judged malignant primarily on the basis of the clinical evidence of local invasion and distant spread. Histologically mitotic figures are rare in these tumours; when present they are only suggestive but not diagnostic of malignancy.

2:2 GENERAL INFORMATION

CBTs fall under a group of tumours called paragangliomas, a family of neoplasms that develop from paraganglion tissues that are themselves chemoreceptor organs distributed throughout the body [11]; therefore these neoplasms can arise from any part of the body that has these tissues. The chemoreceptor organs are of neural crest origin and hence have similar functions and histological appearances [12] Their cells of origin are part of the diffuse neuroendocrine system

(DNES) otherwise formerly called the Amine Precursor Uptake and decarboxylation (APUD) system[12].

The primary cells of the paraganglia system are the chief cells, which are essentially chemoreceptive cells sensitive to hypoxia, changes in pH and fluctuations in blood carbon dioxide concentrations. Therefore being responsive to changes in the chemical environment of the body, it was suggested by Mulligan in 1950 [13] that tumours arising from this system be called chemodectomas; other synonyms for these tumours are glomus tumours, non-chromaffin paragangliomas, receptomas and glomerocytomas [14].

From an anatomical standpoint, paragangliomas can be classified into [15]:-

a) Branchiomeric paragangliomas, e.g. temporal bone, carotid body, subclavian, aortic, pulmonary
b) Intravagal paragangliomas, e.g. upper mediastinal
c) Aorticosympathetic paragangliomas e.g. retroperitoneal
d) Visceral paragangliomas, e.g. pelvic, vagal, mesenteric.

This study is confined to chemodectomas of the carotid body only, otherwise called carotid body tumours (CBT).

2:3 FUNCTIONAL ORGANISATION OF THE CAROTID BODY ANATOMY

2:31 Anatomy

The carotid body is a small ovoid or irregular mass of compact, pinkish tan tissue, measuring 6mm by 4mm by 2mm on average [16]. In its location at the bifurcation of the common carotid artery in the neck, it usually rests against the medical aspect of the adventitia of the vessel [17]. Blood supply is derived from the external carotid artery, and nerve supply which is mainly sensory, from the glossophanyngeal nerve [18].

2:32 Histology [19]

The carotid body consists of nests of "chief" cells surrounded by a fibrous stroma rich in capillaries. The chief cells are large epithelial cells containing copious fine granular cytoplasm and richly supplied with nerve endings specialised to receive chemical stimuli. This histological pattern which is also reflected in the microscopic appearance of CBTs, is best displayed by reticulin stains.

2:33 Embryology:

The carotid body first appears as a condensation of mesoderm cells in the wall of the third arch artery; there are mesoblastic and neural components, with no contribution from pharyngeal ectoderm [4]. The neural components have been thought to arise from the neural crest, the ganglion nodusum of the vagus, and the petrous ganglion of the glossopharngeal nerve. This origin explains the presence in normal carotid body tissue of ganglion cells, nerve fibres, blood vessels and chief cells; it also explains the ability of paranganglia to have an endocrine function [20].

2:34 Physiology

Stimulation of chemoreceptor tissue of the carotid body produces an action potential in the glossopharyngeal nerve leading to [19]:-

a) an increase in the rate, depth and minute volume of respiration
b) an increase in sympathetic outflow leading to tachycardia, hypertension, increased vasoconstrictor tone, adrenaline release and increased cerebral cortical activity.

Stimuli which evoke these responses are hypoxia, acidaemia, hypercabia, increased blood temperature and drugs like cyanides. The actions of the carotid body and carotid sinus are diametrically opposed.

2:4 EPIDEMIOLOGY

2:42 Incidence:

Of all paragangliomas, CBTs are the commonest; however their incidence is very low, with one study quoting 0.012% of all surgical specimens reported in one hospital [21]. Various authors quote that since the first CBT was reported in 1743, over 500 have been reported in literature [8],[9],[19]. Approximately 10% of patients with CBT have either bilateral or multifocal tumours of the chemoreceptor system [14].

2:42 Age:

CBT is generally a disease of the fifth to sixth decades with an average age of occurrence of 45 years. However it may present at any age, with the youngest documented patient being 7 years old [22].

2:43 Sex distribution

The disease has no sex preponderance. Rodriguez-Cuevas et al compared the difference between high-altitude and low-altitude CBT and found an evident female preponderance (8.3:1) in the former compared to the latter (2:1) [23].

2:44 Predisposing Factors:

a) **Environmental influence:-**

Prolonged residence at high altitude has been associated with a high incidence of CBT [6,7]. Gaylis and Mieny et al also found a causal relationship between CBTs and high altitude in a study carried out in South Africa [8]. From these findings, it has been postulated that the increased incidence of CBT among natives living at high altitudes supported the theory that hypoxia induces hyperplasia of chemorective tissue, with a possibility that hypoxia-induced hyperplasia of the carotid body chief cells predisposes to neoplastisc transformation [22].

b) Genetic/Familial Predisposition:-

That CBTs have a genetic predisposition is suggested by the familial occurrence of the neoplasm. It is reported that 30% of all CBTs are familial and in these 10% have a genetic basis; the mode of inheritance is thought to be autosomal dominant [24]. However Khon JS et al [25] in a study on a family spanning three generations showed that the mode of inheritance was not simply autosomal dominant but it appeared to be paternally directed with complete penetrance. From a microgenetic point of view Wang DG et al [26] in a study on expression of apoptosis-suppressing gene bcl-2 in human CBTs demonstrated that deregulation of apoptosis may be a critical component in the multistep tumourigenesis of CBT and that the expression of oncoprotein bcl-2 may contribute to the generation of these tumours. Additionally the association of familial CBT with bilateral occurrence of the tumour has been demonstrated.

2:5 PATHOLOGY

2:51 Gross:

The CBT is adherent to the medial aspect of the common carotid artery adventitia. As it increases in size the tumour separates the internal and external carotid arteries displacing them laterally. Medial displacement is rare [27].

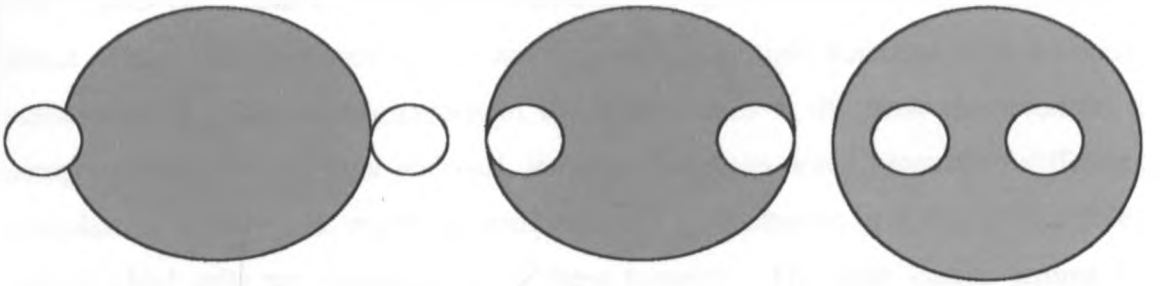
The outer surface may be smooth or irregular with indentations being made by the carotid vessels. Constriction of the vessels is unusual; however compression or total occlusion has been reported [28]. The internal jugular vein, the regional cranial nerves and sympathetic chain are usually displaced by the expanding tumour but may occasionally be encompassed by it. The cut surface has a firm or soft consistency according to the proportion of the fibrous tissue stroma present. According to Shamblin and coworkers [19] increasing degree of circumferential spread around the carotid arteries is associated with increasing tumour adhesion to the vessel wall. It is on this basis that Shamblin has divided the tumour into three groups:-

Group I: - The tumour does not encircle the carotid arteries.

Group II: - The tumour partially encircles the carotid arteries.

Group III: - The tumour completely encircles the carotid arteries.

Figure I: Shamblin Groups:



2:52 Microscopy:

CBT is a neoplasm of the chief cell component of the normal paraganglion with a supporting vascular and connective tissue stroma that contains vessels, nerves, mast cells, lymphoreticular tissue and occasionally small groups of tumour cells. The vascularised connective tissue extends into the tumour in a trabecular manner. Fine reticular fibres and endothelium - lined vessels provide the framework for the nests containing 10 - 20 neoplastic chief cells, the reticular fibres do not however penetrate between the individual cells [29]. The tumours are vascular with blood vessels that vary in size and may either have ill-defined or well-developed walls. The neoplastic chief cells are ovoid, polygonal, rounded or spindle-shaped. LeCompte [20] has classified CBTs into three histological patterns based on the organisation of the chief cells; these are:-

(a) The alveolar pattern: - This is the commonest pattern; it resembles the normal gland architecture with nest cells or "Zellballen" which are usually larger than those seen in the normal gland.

(b) The adenomatous pattern: - This is composed of epithelial sheets of chief cells.

(c) The angiomatous pattern:- This is composed of spindle cells closely applied to capillaries with extensive prominent vascularity.

All the three patterns may be present in one tumour usually with one pattern being predominant.

2:53 Electron Microscopy

The tumour chief cells are polygonal with ovoid or spherical nuclei [30]. The cell bodies are in direct contact with one another and may be attached by tight junctions with desmosome - like substances [31]. The interdigitations of the tumour cells is the most characteristic feature of paragangliomas [31]. A light and dark variation has been noted because of differences in the cytoplasmic density and organelle compactness. Cytoplasmic granules comparable with the normal chief cells are characteristic in these tumours. The inter lobular stroma consists of collagen tissues containing infrequent mast cells, occasional fibroblasts and macrophages. Nerve fibres are rarely seen in the stroma and no axons or synapses have been observed [30]. On electron microscopy these tumours are considered to be truly neoplastic because of their tendency to form monomorphic population of chief cells without sustentacular cells and nerve fibres.

2:6 NATURAL HISTORY

The progression of CBT is characterised by slow and inexorable growth; the symptoms and signs which occur are determined by the alterations of those structures into and around which the tumour grows. Occasionally these tumours grow over time to impressive dimensions without causing any vascular or neurological disturbances. Alternatively the tumour may enlarge into the pharyngeal space, where it presents as a bulge in the tonsil area causing dysphagia.

Despite divergence in opinion regarding the incidence of malignancy [8] presently the tumours are judged malignant primarily on the basis of the clinical evidence of local invasion and distant spread. The common sites of metastasis are lungs, bones, abdominal wall and liver; metastasis to many other sites has been reported. Generally however the incidence of metastasis is low, being estimated at 2-7% of all reported CBTs [19]. CBTs will recur locally if incompletely excised whence they may demonstrate an aggressive local behaviour [1].

DIAGNOSIS

The diagnosis of CBT is made clinically and confirmed radiologically. One must keep in mind the possibility of this diagnosis and consider all single neck masses occurring at the angle of the mandible to be CBT until proved otherwise. Once the diagnosis has been made the tumour's functional status is assessed both clinically and biochemically [1].

2:71 Symptoms:

Presentation of CBT ranges from the asymptomatic cases picked on routine head and neck examination, to dramatic symptoms of rapidly enlarging neck mass causing dyphagia, neck pain or stridor. The commonest complaint though is of a painless neck lump in the region of the angle of the mandible, of variable duration ranging from months to years, with a mean of four years [22]. One patient is reputed to have presented with a neck mass after 47 years [19]. Due to lack of anatomic restriction to growth in the neck CBTs may grow to an enormous size before producing symptoms most of which arise due to local extension into surrounding structures i.e. cranial nerves, sympathetic chain, oropharynx and the skull base. The possible symptoms are summarised below:-

Possible symptoms of CBT

▪ Asymptomatic	▪ Syncopal attacks
▪ Neck pain	▪ Hearing loss
▪ Neck discomfort	▪ Tinnitus
▪ Neck mass	▪ Stridor
▪ Voice hoarseness	
▪ Tongue weakness	
▪ Dysphagia	
▪ Headache	
▪ Dizziness	

2:72 Signs

The commonest sign is usually a single neck mass of variable size, occurring at the level of the carotid bifurcation. With increasing size the tumour may present as a parapharyngeal space mass [32]. The mass may be firm, soft or compressible and is mobile horizontally but not vertically. Davidge - Pitts and Pantanowitz [1] in their study of this tumour found this mobility to be of great diagnostic value in their patients; furthermore they found that mobility was dependent on the tumour size, with small lesions showing great mobility and bigger lesions being less mobile. However this feature is not pathognomonic of CBT.

Pulsation is a universal finding in all CBTs; this is thought to arise from the carotid vessels or less commonly from the vascularity of the tumour itself. Bruits are uncommon as the tumour seldom causes turbulent carotid blood flow. If present the bruits will disappear with carotid compression [33]. Digital compression of the tumour causes shrinkage in all cases, though less commonly so in the large tumours.

The incidence of cranial nerve palsies has been estimated at 20% with the commonest nerves involved being the vagus, hypoglossal and the sympathetic chain. Rarely the glossopharyngeal and the accessory nerves may be involved too. Other commoner signs of CBT are summarised below:

Signs of CBT

- Cervical mass
- Parapharyngeal space mass
- Lateral mobility
- Pulsatility
- Compressibility
- Bruit
- Cranial nerve palsies
- Horner's syndrome

2:73 Differential Diagnosis:

The list below summarises the differential diagnosis of CBT.

Differential diagnosis of CBT[19]:-

- Branchiogenic cyst
- Carotid artery aneurysm
- Tuberculous cervical adenitis
- Lymphoma
- Metastatic carcinoma
- Glomus jugulare tumour

2:8 Radiological Evaluation

Radiology is the mainstay of confirmation of a CBT.

2:81 Plain X-rays:-

Plain x-rays of the base of the skull are done to show a soft tissue mass, displacement of the trachea and larynx, and erosion of the base of the skull; however these features are not consistently demonstrated in all patients with CBT. There being better radiological investigations, plain x-rays are of limited value.

2:82 Angiography:-

It demonstrates a great accuracy in diagnosing CBT. The characteristic appearance of the tumour was first described in 1951[34] and subsequently confirmed by others [35]. It not only confirms the diagnosis but also allows evaluation of coexistent atherosclerotic disease and multicentric cervical paragangliomas. Documentation of a circumscribed vascular blush at the level of the carotid bifurcation, with displacement and separation of the internal and external carotid arteries (splaying of the arteries) is diagnostic. Angiography also demonstrates the anatomic extension, feeder vessels and degree of vascularity of the tumour, all of which are paramount in planning treatment. It also enables the demonstration of deformation of the carotid

arteries by the tumour, whence difficulties in operative excision would be anticipated [37]. Both the left and right carotid systems must be evaluated to rule out multicentric tumours. Lately developed is digital subtraction angiography (DSA) which is a more advanced angiographic technique for CBT evaluation. It gives detailed characteristics of the tumour in terms of soft tissue and vascular composition.

2:83 Doppler ultrasound:

The clinical application of this technique in evaluating CBT has been surpassed by angiography, CT scanning and magnetic resonance imaging (MRI). According to Kapfer X et al [38] the best diagnostic procedure for CBT is a combination of Doppler colour flow imaging ultrasound, computed tomography and selected angiography.

2:84 CT Scanning:-

With this technique CBT appears as a rounded moderately enhancing mass. Dynamic CT Scanning visualises the sequential enhancement of the jugular vein and carotid arteries, and the tumour becomes densely opacified transiently as the intravenous bolus of contrast medium traverses the vascular bed of the tumour. It clearly shows the degree of circumferential spread of the tumour around the carotid vessels. This, combined with its non-invasive nature makes it a relatively superior investigative technique for CBT.

2:85 Magnetic Resonance Imaging (MRI):-

Most, if not all of the information demonstrated by MRI can be derived from CT angiography. In a study on the usefulness of CT and MRI in pre-operative diagnosis of CBT, Sagawa T et al concluded that they were useful in decision on the surgical method [39], with MRI being considered suitable for discerning the inner surface of the carotid artery, but could not necessarily reveal tumour invasion of the carotid artery wall. MRI is superior to CT in delineating soft tissue and in detecting minute tumours.

2:9 FUNCTIONAL EVALUATION

The carotid body is a chemoreceptive organ; however its tumours rarely produce an exaggeration of this function [19] and therefore solitary CBTs are not routinely functionally evaluated. However in cases of bilateral or confirmed familial CBT it is prudent to functionally evaluate the tumour, more so to rule out the presence of other functionally active tumours e.g. phaeochomocytomas. This evaluation involves assay of serum and urine catecholamines.

2:10 MANAGEMENT

Treatment modalities for CBT include observation, radiotherapy and surgery [1]. There are no known effective cytotoxics [40].

Observation:

If untreated, CBTs slowly enlarge [41]. This is the rationale behind a non-operative approach in elderly unfit patients. Dramatic acceleration of growth with its consequences and a definite incidence of malignancy must not be overlooked when pursuing this course of management [1]. It is therefore considered wiser to leave a CBT alone in a poor risk patient or one with an advanced tumour than to attempt surgical excision.

Radiotherapy

Initially thought to be radioresistant, CBTs have now been shown to be radiosensitive [14]. Whereas some authors claim beneficial results with this form of therapy, others have recommended it as a primary form of treatment [36]. Treatment with radiotherapy is difficult to justify particularly because complete surgical removal may be curative, and because of the post-irradiation complications which include laryngeal strictures [29], carotid artery and mandibular radionecrosis [32], and complicated subsequent surgical removal [37]. Proponents of radiotherapy recommend it:-

- for tumour that are too large to excise
- as a back-up treatment for those with recurrences
- as an adjuvant to surgery
- very rarely for metastatic disease [40]
- in patients who prefer radiotherapy to surgery
- for poor risk patients, i.e. very old and/or very frail.

Surgical Management

Surgical removal of CBT offers a clinical dilemma in that the arguments for and against aggressive surgical excision are based on the varying opinions on the biological behaviour of the tumour versus morbidity associated with excision [29],[20],[42]. However, many authors [1], [19], [40] recommend the excision of all CBTs provided the patient is a suitable operative risk and the tumour is evaluated as being operable, since waiting would only result in further growth thus increasing the risk of an operation. Conley [43] and Shamblin [19] have outlined the indications for surgical excision; these are:-

a) Histologically malignant but resectable tumour
b) Aggressive growth patterns
c) Small tumors occuring in a patient aged 60 years or less
d) Interference with swallowing and/or breathing and speaking.

Surgical management is oriented towards complete tumour excision, preserving cranial nerve function and ensuring that safety measures, such as the maintenance of cerebral blood flow are taken. These precautions reduce the possibility of neurological deficits developing. Advances in arterial surgery, carotid bypass, pre-operative embolisation, hypothermia and a better understanding of the physiology of cerebral blood flow have led to a drastic drop in the morbidity and mortality from CBT surgery [44].

The type of surgical procedure used is influenced by the Shamblin category of the tumour [19]. Shamblin group 1 is managed by a complete excision of the tumour carried out in the sub-adventitial plane.

In groups II and III it is recommended that prophylactic carotid shunting be employed. In the former group this facilitates the repair of a damaged vessel after the tumour has been dissected out. In group III tumours atrophic media and adventitial dictate that the tumour must be excised en-bloc together with the carotid arteries, followed by primary carotid artery grafting. In the few instances where extension of the tumour to the base of the skull leaves insufficient internal carotid artery for anastomosis, no attempt should be made to resect this tumour since in such cases arterial reconstruction is not feasible [32].

Pre-operative Preparation:

a) Laboratory work-up

- Full Haemogram; urea and electrolytes; these should be within normal range. If deranged, they should be corrected.
- Group and crossmatch:- At least four units of whole blood should be availed for possible transfusion

b) Direct and indirect laryngoscopy:-

This is done to assess ninth cranial nerve involvement and to look for possible pharyngeal involvement by upward extension of the tumour.

c) Pre-operative embolisation:-

Angiographic embolisation of tumour-supplying arteries - commonly from the external carotid and occasionally from the vertebral or other arteries - significantly reduces intraoperative haemorrhage thus improving operability. Koch G, Klein GE et al [45] in a review of 13 patients with CBTs that had been declared inoperable by virtue of upward extension to the skull base, employed this technique and managed to achieve curative surgical resection. Once the tumour has been successfully devascularised, surgery should be performed within 2-3 days to avoid revascularisation.

THE OPERATION [46]

a) Anaesthesia:-

The operation is done under general anaesthesia; nasal intubation improves access to a tumour extending up under the mandible into the base of the skull. Central venous pressure and arterial blood gases are constantly monitored.

b) The Surgeon:

Ideally the operation is best performed by a qualified and experienced vascular surgeon.

c) The Procedure:-

Under general anaesthesia the patient's head is turned away from the side of the lesion and placed on a rubber ring. Both the operation site and the groin are prepared and draped, the latter in case a length of long saphenous vein is required. The incision is made from the mastoid tip continuing inferiorly along the anterior border of the sternocleidomastoid muscle and curving medially at the level of the carotid bifurcation to end just before the midline. Adequate exposure of the carotid system is essential. The common, internal and external carotid arteries are each exposed and controlled with vascular clamps beyond the tumour. Some surgeons advocate dividing the external carotid artery near its origin as soon as possible to reduce vascularity of the tumour and improve access, whereas others advocate clamping of the artery at this point; heparin is not required prior to this. A shunt is inserted before dissection of the tumour begins in groups II and III patients only.

Gaylis and Mieny et al [8] advocate the loose periadventitial connective tissue plane as the safer one along which cleavage should be effected. They advise avoidance of the subadventitial plane [1] since cleavage along it exposes the media which is friable, easily damaged and holds sutures poorly. Once the tumour has been adequately exposed and the shunt inserted, the tumour dissection is effected until the tumour is freed from the internal carotid artery initially.

The tumour feeding vessel is identified and ligated. The external carotid artery feeders are treated similarly, or the main artery transected and ligated as previously outlined. Throughout the dissection every attempt should be made to get a clear view of the cranial nerves as they run across the deep aspect of the upper part of the tumour. Good visualisation and haemostasis enable clear identification and preservation of the cranial nerves especially the vagus and hypoglossal. However greatly they are involved the tumour can often but not always be shelled off them.

Group III tumours usually dictate that the internal carotid artery be excised due to extensive involvement by tumour, in which case arterial grafting using long saphenous vein or dacron graft will be carried out. Large locally invasive tumours may necessitate resection of the regional lymphnode and if unavoidable regional cranial nerves [47]. Once the tumour has been removed the carotid arteries are clampled, the shunt removed and the shunt insertion sites closed. The final sutures are placed and tied, ensuring meticulous haemostasis, and wound closure over a vacuum drain, which is left in place for 24-48 hours.

Post-operative care:

This aims at ensuring smooth post-operative recuperation of the patient, identification and where possible appropriate correction of complications. It entails:-

a) Post-operative monitoring of blood pressure, central venous pressure pulse rate and cerebral blood flow especially where the internal carotid artery has been reconstructed.
b) Monitoring for possible haemorrhage
c) Monitoring for central and peripheral neurological deficits e.g. ninth, tenth and twelveth cranial nerve palsies and cervical sympathetic nerve palsy.

2:11 COMPLICATIONS:

a) Nerve Palsies:-

The most common nerves to be injured are the vagus and hypoglossal [48]. The only way to avoid injury is to positively identify them before proceeding with tumour excision. The entire course of the nerve from the base of the skull to beyond the limits of the tumour must be followed. Most injuries are transcient, improving after a few weeks. The ninth and eleventh cranial nerves may also be injured.

b) Horner's syndrome due to cervical sympathetic ganglia damage
c) Hoarseness of voice due to recurrent laryngeal nerve damage
d) Cerebrovascular accidents (CVAs)
e) Haemorrhage in the immediate post-operative period.

2:12 FOLLOW-UP

For patients who do not undergo surgery by virtue of age, poor surgical risk or extensive tumour involvement, follow-up is aimed at identifying evidence of metastasis, life-threatening symptoms and signs, whence other interventions eg radiotherapy would be employed. For post-operative patients follow-up aims at identifying recurrence, contralateral occurrence of tumour (whence familial trait should be ruled out), evidence of metastasis and occurrence of complications. At Kenyatta National Hospital these patients are followed up in the Cardiothoracic Clinic where they are reviewed by specialist vascular surgeons in conjunction with trainee surgeons.

STATEMENT OF RESEARCH PROBLEM:

3:1 RESEARCH PROBLEM

Carotid Body Tumour is a frequently misdiagnosed and hence mismanaged condition in our set-up; this is true especially in the peripheral health establishments where most of these patients present before being referred to Kenyatta National Hospital. The main reasons for this scenario are that the condition is rare and when it occurs it presents as an innocent i.e. asymptomatic mass in the neck which is easily branded a lymphnode, or a lipoma; secondly there lacks a high index of suspicion among clinicians to adequately make an impression of carotid body tumour. As a result of the foregoing most patients presenting at Kenyatta National Hospital as referrals have either had attempts at aspiration, biopsy or even excision of the mass, all of which are dangerous manouvres in as far as management of CBTs is concerned.

3:2 JUSTIFICATION OF THE STUDY:

This study is based on the fact that absolutely no local data exists on the management of CBT at KNH. This is inspite of the fact that on average there are 2 or more new cases of CBT diagnosed annually. Secondly there occur no clear-cut guidelines to assist both medical and paramedical personnel in having a high index of suspicion that a solitary neck mass could indeed be a CBT and hence the need for prompt referral. Presently the expertise and diagnostic and surgical equipment for competent management of CBTs are based in Nairobi, either in KNH or probably in other major private hospitals. It therefore behooves us as surgical practitioners to be able to diagnose CBT beyond any shadow of doubt, and to ensure its proper management by the relevant experts, i.e. cardiovascular surgeons, in order to pre-empt both the morbidity and mortality that is invariably associated with the misdiagnosis and eventual mismanagement of this rare albeit manageable condition. From this study it is hoped that a management protocol for CBT in our set up shall be formulated.

3:3 STUDY OBJECTIVES

3:3 BROAD OBJECTIVE:

To review presentation and management of Carotid Body Tumour at Kenyatta National Hospital(KNH).

3.32 SPECIFIC OBJECTIVE

1. To highlight the epidemiological pattern of CBT in our set up.
2. To identify the presenting clinical features of CBT as seen at KNH.
3. To document the magnitude of CBT as a surgical problem in KNH.
4. To review the diagnostic workup for CBT as done at KNH.
5. To review the treatment of CBT and its outcome at KNH.
6. To establish a management protocol for CBT at KNH for future guidance and reference.

4.0 METHODOLOGY

4:1 STUDY DESIGN:

This was a retrospective study covering a period of eleven years from January 1990 to December 2000.

4:2 STUDY AREA:

Kenyatta National Hospital; Thoracic and Vascular Surgery Unit.

4:3 STUDY POPULATION

All patients diagnosed to have carotid body tumour (CBT) during the study period.

4:4 SAMPLING

4:41 Sample Unit

Thoracic and vascular surgery unit - Kenyatta National Hospital

4:42 Sample Frame

Registry and Records Department - Kenyatta National Hospital

4:43 Sample size

Same as the study population

4:44 Sample Procedure

Purposeful, i.e. the target was patients with CBT only.

4:5 DATA COLLECTION, PROCESSING AND ANALYSIS

4:51 Instruments

Patients' medical records (files)
Operation registers
Radiological films

4.52 Personnel

The investigator himself.

4:53 Processing and Analysis:

The data was derived from the patients' records using a data collection form. It was carefully monitored and entered into a compatible computer.

Analysis was carried out using SPSS/PC + to derive descriptive characteristics and frequency distribution of the study population.

4:6 MINIMISATION OF ERRORS AND BIASES:

This was achieved by adoption of inclusion and exclusion criteria

The inclusion criteria was:-

a) Patients with a histological diagnosis of CBT.
b) Patients with an angiographic diagnosis of CBT.
c) Patients with recurrence of previously treated CBT.

The exclusion criteria was:-

a) Patients with neck masses that did not conform to an angiographic or histologic diagnosis of CBT.

b) Patients lost to follow-up after diagnosis of CBT.

c) Patients with incomplete or unavailable records at KNH.

4:7 ETHICAL CONSIDERATIONS:

The study proposal was submitted to the Kenyatta National Hospital Ethics and Research Committee for approval. All the data concerning the patients under study was treated in a strictly confidential manner. Patient's identity was not revealed to unauthorised persons.

5. PRESENTATION OF RESULTS

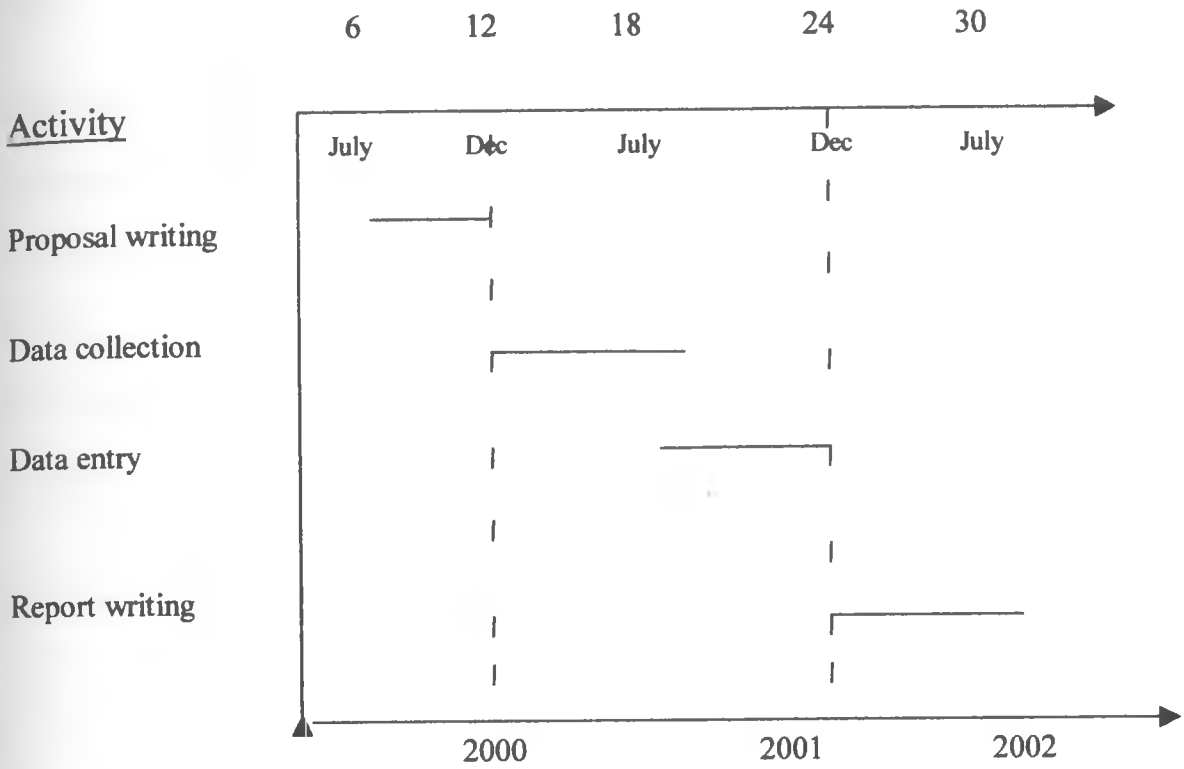
The data was presented in tabular and graphical forms, with use of bar and pie charts where necessary.

6. LIMITATIONS OF THE STUDY

1. The rarity of the condition under study dictated that the sample size had to be small.
2. The retrospective nature of the study necessitated derivation of data from patient's medical records which may have been incomplete and/or inaccurate.
3. The radiological investigations for this condition are expensive and hence some patients were not able to afford them.
4. The follow-up of patients after surgery was erratic and hence it was not possible to document all the post-surgical sequelae.

7.0 ORGANISATION OF THE STUDY (WORKPLAN)

TIME FRAME (MONTH)



8. COST ESTIMATE (BUDGET)

ITEM	COST (KSHS)
Data Collection	15,000.00
Computer analysis	15,000.00
Stationery	5,000.00
Typing	5,000.00
Final compilation	20.000.00
TOTAL	**60.000.00**

The source of funding was the investigator's personal resources.

RESULTS

Between January 1990 and December 2000, a total of 30 patients with CBT were seen and treated at Kenyatta National Hospital. The Management details are summarised in the results below:-

Table 1: Year of Presentation of Patients with CBT to KNH (n=30)

YEAR	FREQUENCY	%
1990.	3	10.0
1991.	4	13.3
1992.	1	3.3
1993.	2	6.7
1994.	2	6.7
1995.	2	6.7
1996.	7	23.3
1997.	3	10.0
1998.	1	3.3
1999.	3	10.0
2000.	2	6.7
TOTAL	**30**	**100**

From the above the annual prevalence of CBT at KNH is 2.72.

Figure 1: Year of Presentation of patients with CBT to KNH

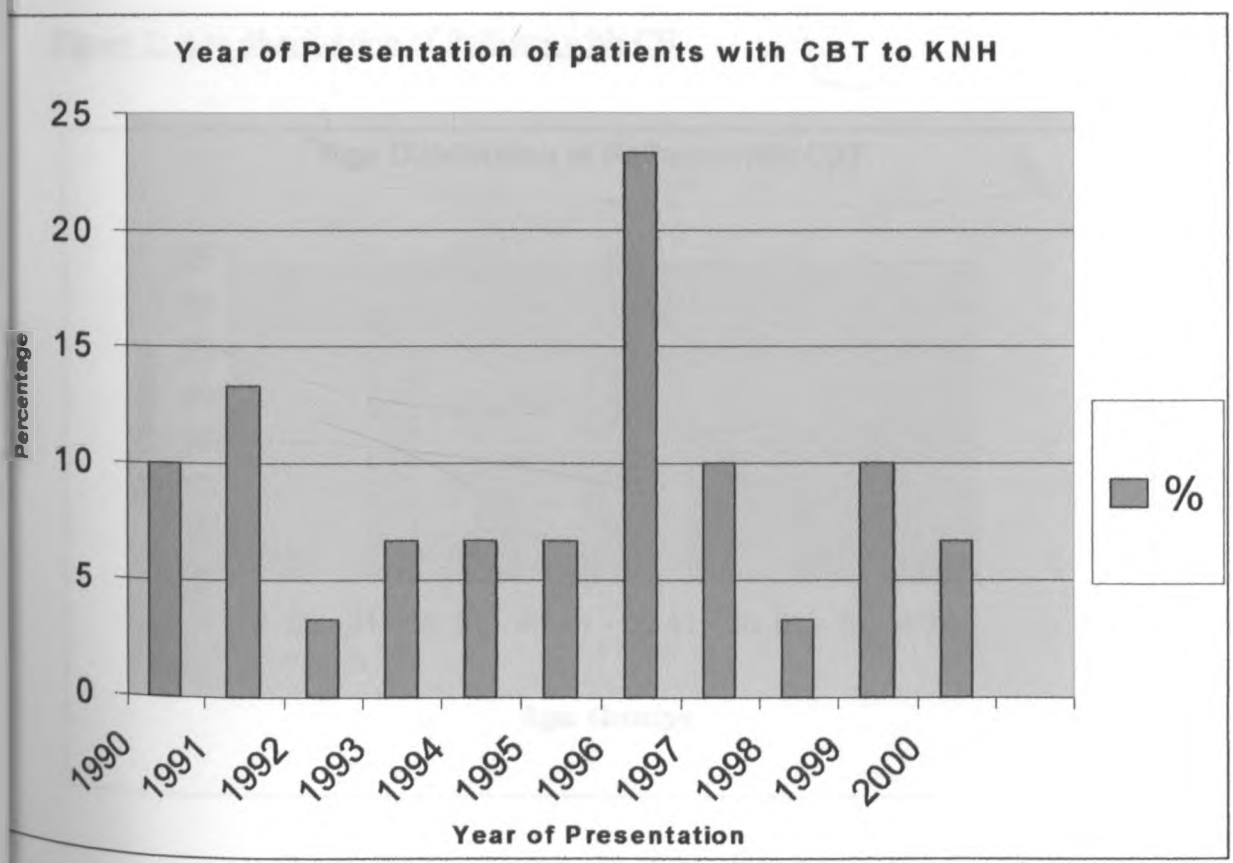


TABLE 2: AGE DISTRIBUTION OF PATIENTS WITH CBT

AGE GROUP (YRS)	FREQUENCY	%
11 - 20	1	3.3
21 - 30	9	30.0
31 - 40	4	13.3
41 - 50	7	23.4
51 - 60	5	16.7
61 - 70	3	10.0
> 70	1	3.3
TOTAL	**30**	**100.0**

Age Range (years) - 20 - 96

Mean Age (years) - 43.3

Mode (years) - 45.0

Table 2 shows age distribution of patients with CBT. The age range was 20 - 96 years with a mean age of 43.3 years and a mode of 45 years. The youngest patient was aged 20 years and the oldest 96 years. Majority of the patients were in the 21 - 60 years age range (83.4%). The peak occurrence was in the 3rd decade i.e. 9 patients (30%).

Figure 2: Age distribution of Patients with CB

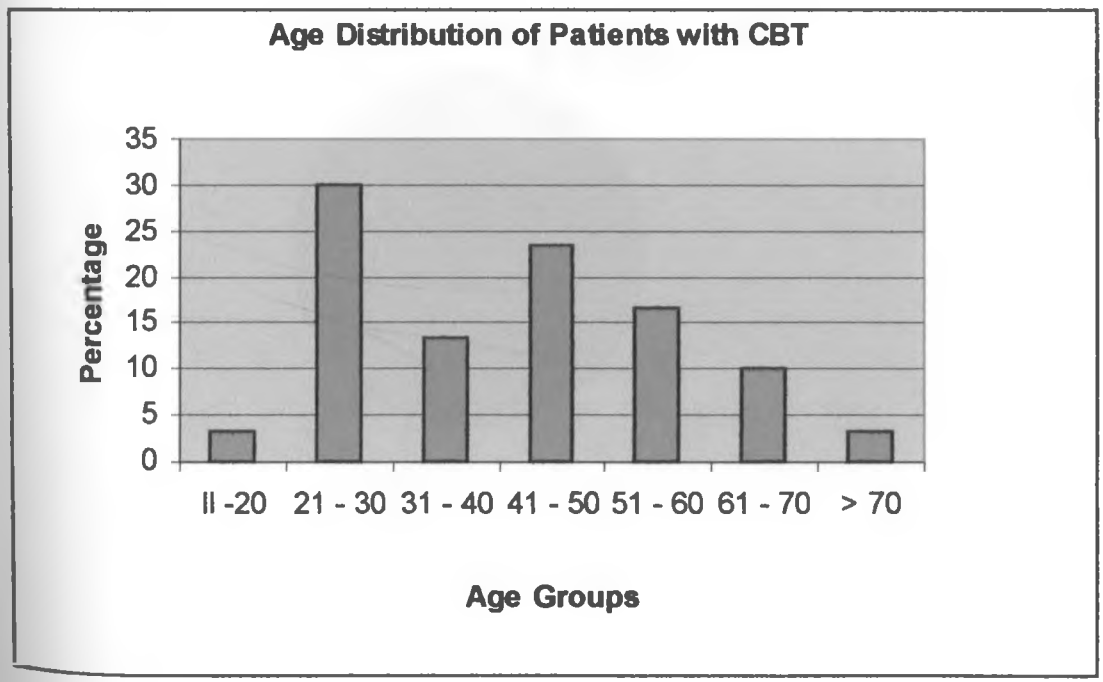
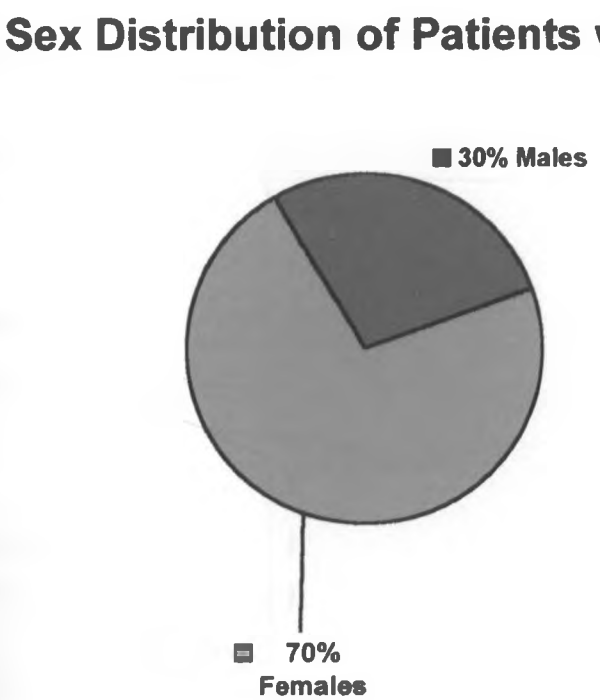


Table 3 :Sex Distribution Of Patients With CBT (N=30)

Sex	Frequency	%
Female	21	70%
Male	9	30%
Total	30	100%

Table:3 shows the sex distribution of the 30 patients with CBT. Of these 21 (70%) were female and 9(30%) male. The female to male ratio (F:M) was 2.33:1 or 7:3

Figure 3: Sex Distribution Of Patients With CBT



Geographical Distribution (Home District) of patients with CBT

Of the 64 districts in the whole country, CBT was found in only 16(25%) . The distribution was as indicated in table 4 below:

Table: 4:

Geographical Distribution (Home District) of patients with CBT

District	Frequency	%
Baringo	1	3.3
Kakamega	1	3.3
Kiambu	6	20.0
Kisii	6	20.0
Machakos	2	6.7
Meru	1	3.3
Muranga	2	6.7
Nairobi	2	6.7
Nakuru	1	3.3
Nyamira	1	3.3
Nyandarua	1	3.3
Nyeri	2	6.7
Siaya	1	3.3
Taita Taveta	1	3.3
Tranz-Nzoia	1	3.3
Vihiga	1	3.3
Total	**30**	**99.8**

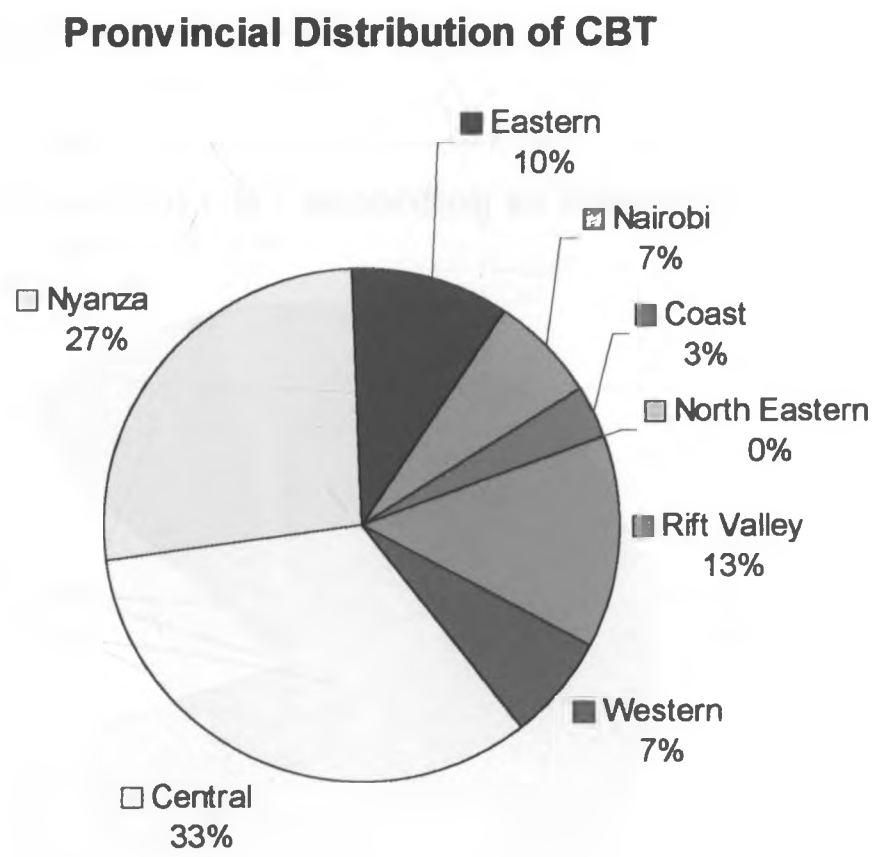
The Provincial distribution of CBT was as indicated hereunder (Table 5).

Table 5:

Distribution of CBT according to Provinces.

Province	Frequency	%
Rift Valley	4	13.3
Western	2	6.7
Central	10	33.3
Nyanza	8	26.7
Eastern	3	10.0
Nairobi	2	6.7
Coast	1	3.3
North Eastern	0	0
Total	30	100.0

Fig 4: Provincial Distribution of CBT



Central Province. Nyanza Province had the second highest number of patients with CBT (26.7%). Only one patient from Coast Province was treated for CBT whereas none came from North Eastern Province.

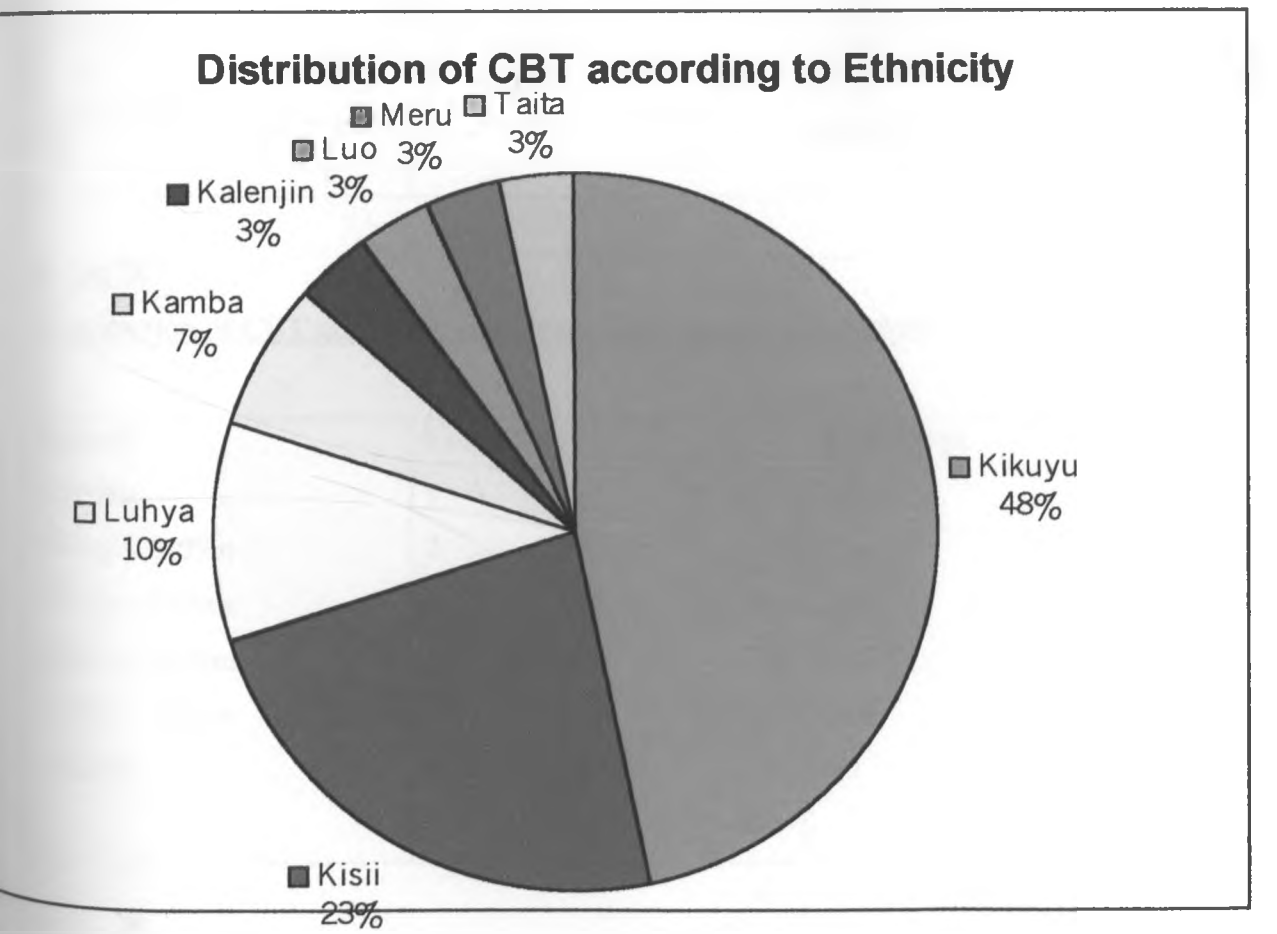
Table 6:

Distribution of CBT According to Ethnicity (Tribe)

Tribe	Frequency	%
Kikuyu	14	46.7
Kisii	7	23.3
Luhya	3	10.0
Kamba	2	6.7
Kalenjin	1	3.3
Luo	1	3.3
Meru	1	3.3
Taita	1	3.3
Total	30	99.9

Of the 42 ethnic groups in Kenya, only 8 were afflicted by CBT . The highest number was represented by the Kikuyus followed by the Kisiis.

Fig.5:



Distribution of CBT according to altitude (metres above sea level)

It is known that there is a high prevalence of CBT at high altitude. The results in this set-up were not in keeping with this knowledge.

Table 7A:

Distribution of CBT according to altitude.

District	Frequency	Approximate Altitude (metres ASL)
Muranga	2	1400m
Kakamega	1	1500m
Siaya	1	1500m
Taita Taveta	1	834m
Nairobi	2	1700m
Machakos	2	1750m
Baringo	1	1780m
Kiambu	6	1800m
Vihiga	1	1800m
Nyeri	2	2000m
Nyamira	1	2000m
Kisii	6	2000m
Nakuru	1	2100m
Meru	1	2500m
Nyandarua	1	2500m
Trans-Nzoia	1	2850m

Table 7B:

Distribution of CBT according to altitude (metres above sea level)

Altitude	Frequency	Percentage
<1000m	1	3.3
1000m - 1499m	2	6.7
1500m - 1999m	14	46.7
2000m - 2499m	10	33.3
2500m - 2999m	3	10.0
>3000m	0	0.0

Fig. 6:

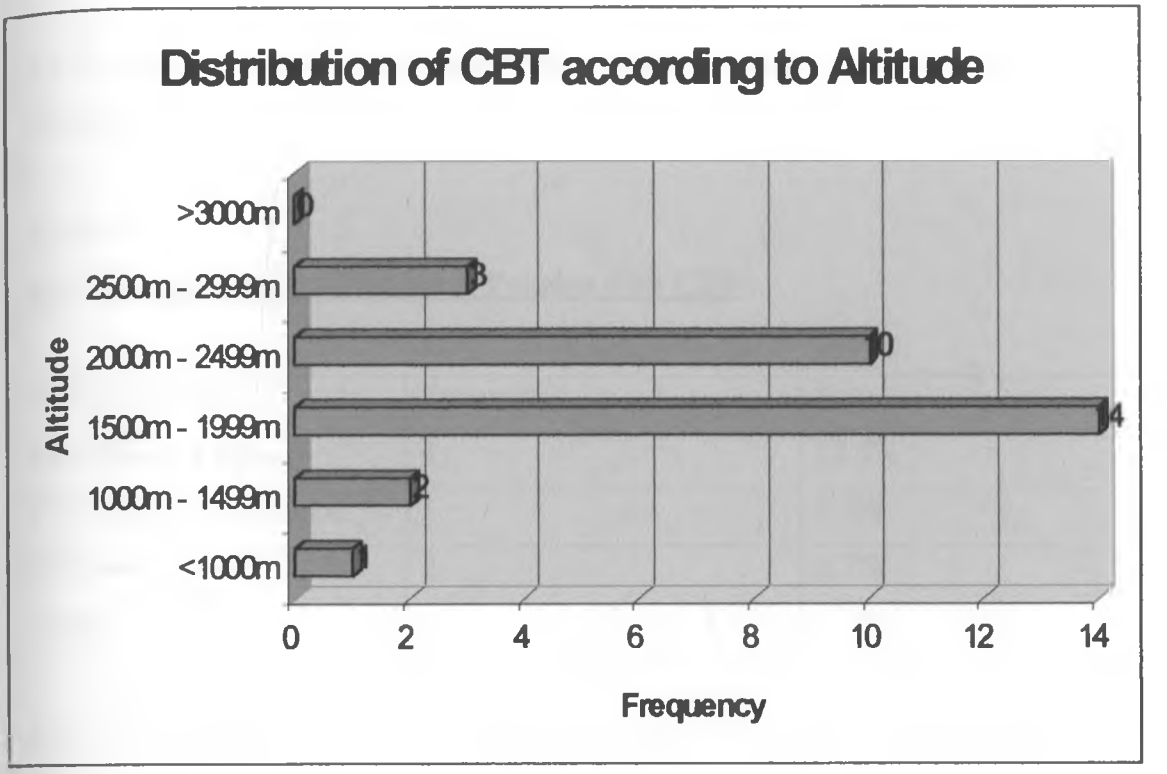


Table 8:

Symptoms of patients with CBT

Symptom	No. of Patients	%
Lump(s) in neck	30	100
Neck pain	14	46.7
Neck discomfort	13	43.3
Headache	11	36.7
Hoarseness of voice	6	20.0
Dysphagia	5	16.7
Dizziness	5	16.7
Others	4	13.3

All the patients presented with a lump in the neck. The next commonest symptoms were neck pain (46.7%) and neck discomfort 43.3%.. Among other symptoms that

13.3% patients presented with were ear pain (otalgia), vertigo, facial weakness and pulsatility of the lump.

Table 9:

Relevant Past Medical History of Patients with CBT

Medical history	Frequency	%
Previous neck surgery	4	13.3%
Previous neck irradiation	1	3.3%
Previous treatment for cancer	2	6.7%

Four patients gave a history of previous neck surgery. Of these 3 had undergone thyroidectomy and one had an incisional biopsy for a neck mass, which turned out to be a carotid body tumour.

Two patients had received treatment for cancer, one was on tamoxifen for treatment of breast cancer, the other had undergone subcapsular orchidectomy for treatment of prostate cancer.

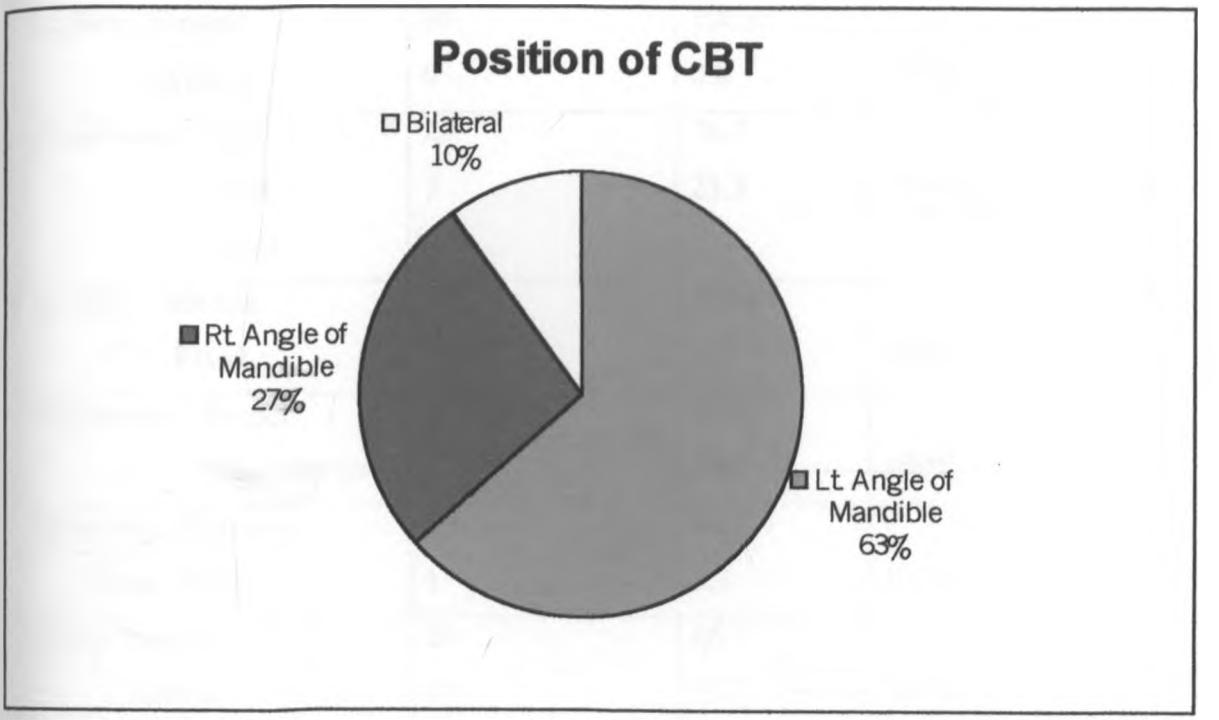
Characteristics of the CBTs

Table 10: Position of the CBT:

Position	Frequency	%
Lt. angle of mandible	19	63.3
Rt. angle of mandible	8	26.7
Bilateral	3	10.0
Total	30	100.0

The majority of CBTs were located at the angle of the mandible on the left (63.3%). Ten percent of patients had bilateral tumours, all of whom were females.

Fig. 7: Position of CBT



Size of the tumour as seen at histology

The tumours ranged in size from the smallest (2cmX 2cm X 4cm) to the largest (15cm X 15cm X 15cm) on average.

Table 11: Other Characteristics of the CBT

Characteristic	Frequency	%	Total
Shape - Spherical	15	50	
- Ovoid	10	33.3	100%
- Pear shaped	5	16.7	
Surface - Smooth	30	100.0	
- Irregular	0	0.0	100%
Consistency - Soft	23	76.7	
- Firm	7	23.3	100%
- Hard	0	0.0	
Mobility - Mobile	30	100.0	
- Fixed	0	0.0	100%
Tenderness - Tender	3	10.0	
- Non - tender	27	90.0	100%
Pulsatility - Pulsatile	29	96.7	
Non - Pulsatile	1	3.3	100%
Bruit - Present	20	66.7	
- Absent	10	33.3	100%

Table 12: Distant Signs in Patient with CBT

	Frequency	%
Parapharyngeal space mass	4	13.3
Nineth cranial nerve palsy	2	6.7
Seventh cranial nerve palsy	1	3.3
Twelveth cranial nerve palsy	1	3.3
CNS(Cord compression T9)	1	3.3
	9	30.0

The majority of patients (70%) had local signs; of the 30% with distant signs, 4 (13.3%) had parapharyngeal mass confirmed by laryngoscopy. The patients with nineth and twelveth cranial nerve palsies had bilateral CBT.

Table 13: Radiological Assessment of CBT (n=30)

Investigation	Frequency	%
Plain X - Rays (base of skull)	3	10
Ultrasound	18	60
Angiography	21	70
CT Scan	10	33.3
MRI	0	0
None	1	3.3

Fig. 8: Radiologic Investigations of Patients with CBT

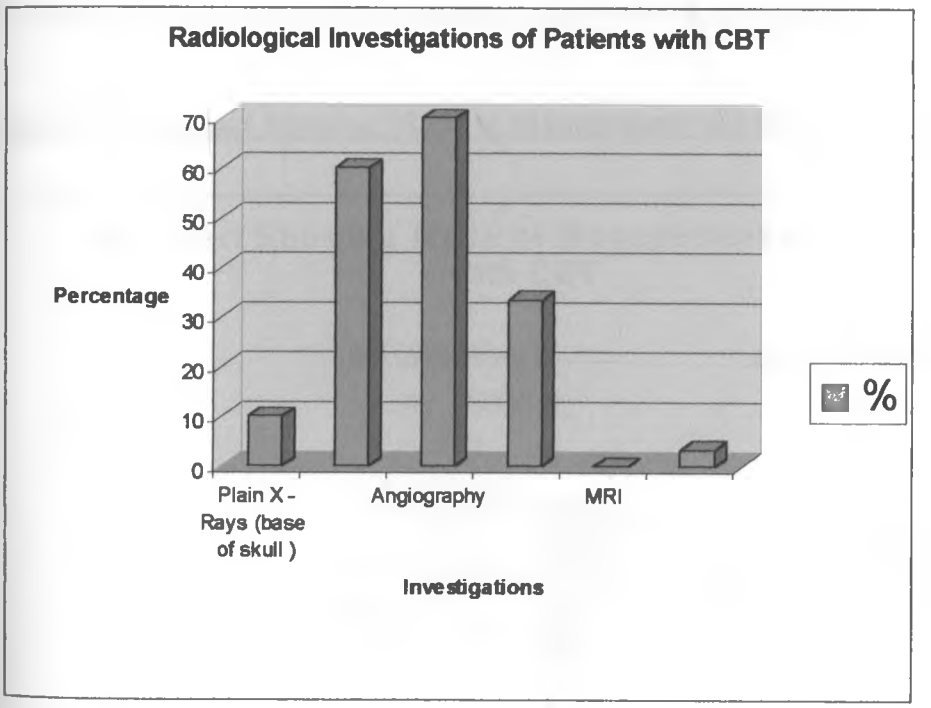


Table 14: Definitive Management

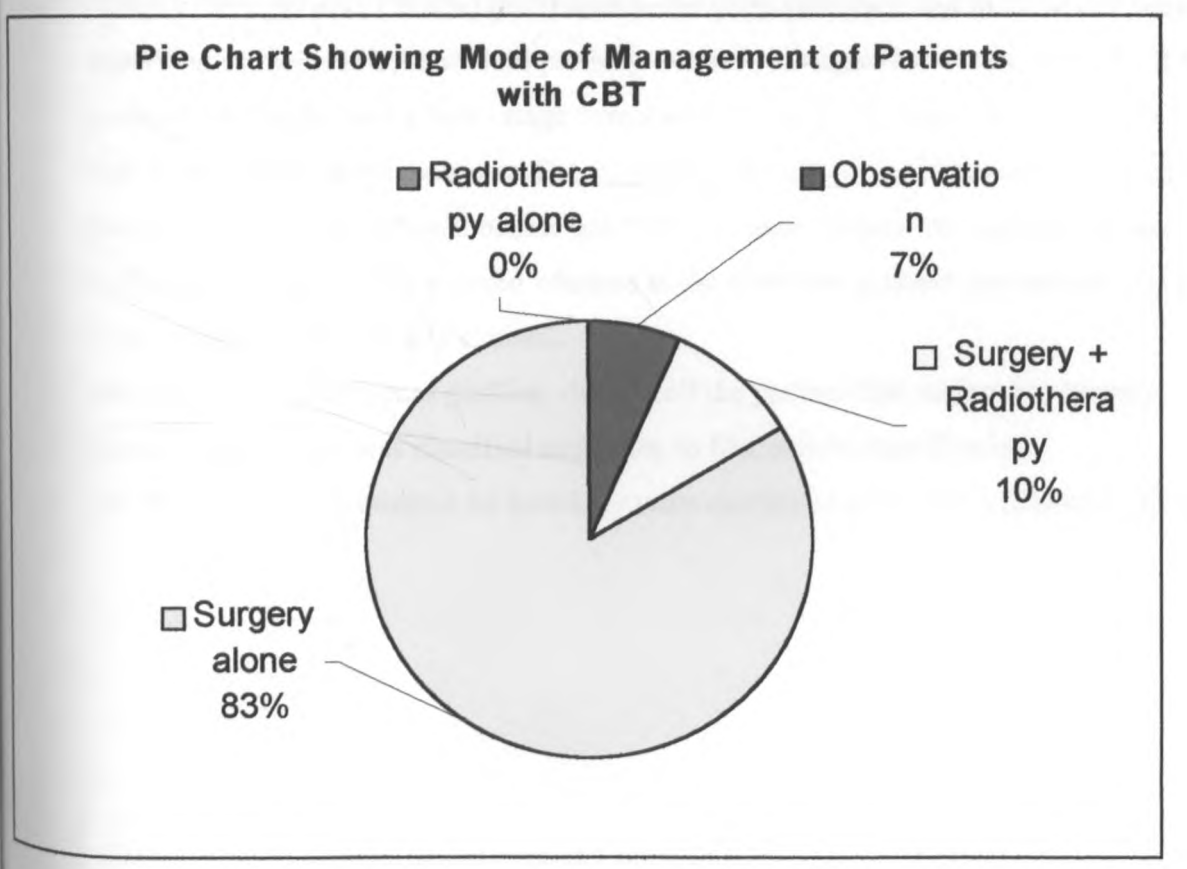
Table showing mode of management for patients with CBT

Mode of Management	Frequency	%
Radiotherapy alone	0	0
Observation	2	6.7
Surgery + Radiotherapy	3	10.0
Surgery alone	25	83.3
Total	30	100

No patients was treated with radiotherapy alone. Two (2) patients who had angiographic diagnosis of CBT declined surgery and hence were followed up in the clinic.

In total 28 patients (93.3%) were operated on; of these two received radiotherapy as an adjuvant to surgery in view of the incompleteness of the excision of tumour.

Figure 9: Pie Chart Showing Mode of Management of Patients with CBT



Pre-Operative Intervention

Not a single patient in the study underwent preoperative irradiation of the tumour or embolisation of the tumour vasculature.

Operative Intervention

Table 15:

	Frequency	%
One Stage Operation		
- Inoperable	1	3.3
- Partial Excision	4	13.3
- Total Excision	18	60.0
Two Stage Operation		
- For Large CBT	2	6.7
- For Bilateral CBT	3	10.0
Total	28	93.3

- Twenty three patients (76.6%) underwent a one stage operation; out of these one tumour was described as inoperable, four were partially excised and eighteen totally excised. Of the five patients who underwent a two - stage operation two were because of large tumours which had to be excised in two sessions. The remaining three had bilateral tumours each of which had to be excised on different occassions. Of these patients bilateral tumours, in one patient both tumours were totally excised whereas in the other two patients one tumour was partially excised and the other totally excised.
- Neither was carotid bypass grafting done in all the patients that underwent surgery.
- Not a single tumour was classified according to Shamblin's classification.
- All the specimens submitted for histology were confirmed to be CBTs (chemodectoma).

Outcome of Surgery & Complications.

Table 16: Outcome of Surgery:

Outcome	Frequency	%
Cured	18	60
Recurred	10	33.3
Died	1	3.3

Of the 28 patients operated on, 18(60%) were presumed to have been cured since on subsequent follow - up there was no clinical evidence of recurrence.

Ten (33.3%) of the patients operated on exhibited recurrence of tumour on subsequent follow - up. Of these one died during re - operation of a recurrence.

Table 17: Complications of Surgery

Complication	Frequency	%
Hoarseness of voice	15	50
Nineth Cranial nerve palsy	9	30
Twelveth cranial nerve palsy	9	30
Tenth cranial nerve palsy	3	10
Dysphagia	2	6.7
Eleventh cranial nerve palsy	1	3.3
Cerebrovascular accident	1	3.3

The commonest complication, occurring in 50% of all the patients under study was hoarseness of voice.

Palsy of the nineth and twelveth cranial nerves occurred in an equal number of patients; in each case nine patients were affected..

The least common complications were eleventh cranial nerve palsy and cerebrovascular accident each occurring in 3.3% of the study population.

DISCUSSION

This is the first study on carotid body tumours to be carried out at Kenyatta National Hospital. As such most, if not all, the results are novel and thus there are no previous results with which to compare the findings.

Prevalence:

Concerning the study population that met the inclusion criteria of the study, only 30 patients were seen during an eleven year period, giving an annual prevalence of 2.72.patients. This is in keeping with the rarity of the tumour worldwide, [8],[9],[19]. Most patients were seen in 1996 (table 1) This was incidental, since the trend was not maintained in other years of the study period.

Age Distribution

In this study the youngest patient was aged 20years and the oldest 96years. The mean age of presentation was 43.3 years and the mode 45. The majority of patients (83.4 %) were in the age range 21-60 years with a peak occurrence of the disease in the third decade (30%). The disease may occur at any age, even though it is commoner in the fifth to sixth decades; [22] in this study it occurred maximally in the third decade; similarly the average age of occurrence is quoted as 45years which concurs with the mean age of 43.3years in the study . The very low incidence of the disease above 70years in the study (3.3%only) is inexplicable in the present context, but may be multifactional.

Sex Distribution.

The female: male ratio of CBT in the study was 2.33:1 or 7:3. In the study more female patients (70%) were seen than males (30%) and hence the above finding is not significant. That high altitude CBT is commoner in female than in males as demonstrated by Rodriguez - Cuevas et-al, [23] could not be established in this study.

Predisposing Factors.

The distribution of CBT according to altitude was not significant in this study. The majority of patients (14, ie 46.7%) came from middle -altitude regions (1500-1999m above sea level). Infact only 10% of patients came from high altitude region. The altitudes quoted by Arias Stella [6] and Saldanha and associates [7] as being significant predisposing factors are over 7000ft above sea level. It thus shows that from the local scenario, either the altitudes considered are not an important predisposing factor, or they are not high enough to influence neoplastic transformation of the carotid body.

Of all the patients included in the study, none gave a positive family history of CBT. The same applied to the 3 patients (10%) who had bilateral tumours, which are known to run a familial course. Further studies are needed to establish the familial nature or otherwise of these bilateral tumours in our setup.

A total of 7 patients (23.3%) gave a relevant past medical history (Table 9). Four had undergone previous neck surgery, one had undergone previous neck irradiation and two had received treatment for cancer 6.7% . It is unlikely that the previous neck surgery predisposed to development of CBT in these patients; however there exists a possibility of the previous neck irradiation having contributed to development of CBT in the one patient (3.3%) . This however could not be confirmed. The two patients who had received treatment for breast cancer (with tamoxifen) and prostate cancer (orchidectomy) are unlikely to have been highly predisposed to development of CBT by virtue of their treatment.

Symptoms and signs

All the patients seen complained of a lump in the neck. In the majority though (53.3%) the lump was painless whereas in the remaining ten (46.7%) it was painful. Forty three percent of patients complained of neck discomfort . Interestingly not a single patients lump was discovered incidentally in routine head and neck examination, all having been referred with the specific complaint to a lump in the neck. The other common symptoms were headache (36.7%) and

hoarseness of voice. The pain and discomfort are caused by the pressure effect of the CBT on surrounding

structures in its region of growth e.g the blood vessels and the nerves. Hoarseness of voice is due to compression of the plarynx by the growing CBT, causing pressure or paralysis of the vocal cords. One patient complained of pulsatility of the lump.

Nineteen of the CBTs were located on the left angle of the mandible eight ,(26.7%) on the right and three were bilateral; whether the predilection for the left side was incidental or of any statistical significance can only be confirmed by subsequent studies.

All the tumours had a smooth surface and were mobile horizontally but not vertically. Only one patient had a non-pulsatile tumour; the majority (66.7%) of patients had tumours in which a bruit was present. All these findings were consistant with the known characteristics of the CBTs. The most commonly affected cranial nerves were glossopharyngeal and hypoyglossal (Table 17), contrary to what is quoted that the former is the least commonly affected by CBT. [48] Laryngoscopy was done on 13.3% of the patients who had paraphayngeal extension of the tumour.

Radiological assessment.

Plain x-ray of the base of the skull is sometimes requested to assess the upward extent of the tumour, whence the radiological sign is erosion of the base of the skull. With the advent of better and more informative radiological investigations its role in diagnosis has decreased. In this study 10% of patients had this x-ray done, and it was not informative; furthermore they ended up being investigated using other radiological modalities. It therefore just added extra expenses to the patients' hospital bill. Angiography was the investigation requested most (70%) and in all cases it was diagnostic. It showed the characteristic circumscribed vascular blush of the tumour at the level of the carotid bifurcation, with splaying of the vessels .No patient underwent digital subtraction angiography (DSA) during the period of study probably because it was not yet available as an investigative technique. With its introduction at KNH in late 2000 it should become the investigation of choice. Sixty percent of patients had ultrasound done for diagnosis of CBT. However in most of these patients ultrasound had already been done beforehand, after which angiography was done as a confirmatory test. Ultrasound should only be requested in the absence of the other more informative radiological investigation; even then the doppler technique should be preferably requested since it assesses both the soft tissue and vascular characteristics of the tumour. A third of the patients (33.3%) had CT scan of the neck done to diagnose CBT; this was mainly requested to study the extend of the tumours which were relatively large (over 8cm x 8cm x 8cm). In some of the patients angiography had already been undertaken. CT scan should be prescribed for patients with advanced disease who need further characterization of the tumour, or those who cannot withstand the invasiveness of carotid angiography. One patient was operated upon purely on clinical evidence without any radiological back up. For a hospital of KNH magnitude and with the increasing patient awareness and litigation this trend is best avoided.

Definitive Management

Out of the 30 patients enlisted for the study 25 (83.3%) underwent surgery alone. A further 3(10%) underwent surgery and were thereafter irradiated. Two patients (6.7%) declined any mode of therapy and hence were observed. None was treated by radiotherapy alone.

In terms of pre-operative preparation, functional evaluation which involves assay of serum and urine catecholamines to rule out the presence of other functionally active tumours [19] was not done. Even though these patients did not suffer any intra-operative complications during excision of their tumours, nonetheless it is advisable to functionally evaluate them, more so if they have bilateral occurrence of the tumours, since bilateral tumours are known to occur concurrently with other functionally active tumours especially phaeochromocytomas.[19]

Similarly none of the patients underwent pre-operative irradiation or embolisation of the tumour blood supply. Radiotherapy is known to shrink a huge tumour mass, which would otherwise be inoperable, thus rendering it operable [14]. Since it is available it should be used in patients with huge tumours. In this study had preoperative radiotherapy been used to shrink the very large tumours seen in three patients, the rest of the tumour would have been removed in one operation and not in two as was the case.

Pre-operative embolisation of tumour vasculature was not done because it was not requested for. This procedure if requested and done might have benefited the one patient (3.3%) whose tumour was declared inoperable, and the 4 (13.3%) whose tumours were partially excised. Similarly it might have benefited the 2 patients with bilateral tumour in whom only one of the tumours was completely excised and the other partially excised.

Operative Intervention

Twenty three patients (76.6%) were operated on in one sitting whereas 5 (16.7%) were operated on in two sessions because of the tumour being very large- 6.7% or its bilateral occurrence (10 %). In all the patients attempts were made to excise the tumour completely with preservation of

normal anatomy. However not a single patient underwent intraoperative carotid artery bypass grafting.

The reason for this could not be explained but may be attributed to

- surgeon's preference
- lack of appropriate bypass grafting material
- lack of intraoperative indication for the same.

Other series have highlighted the merits of application of these techniques. A possible explanation to the non-usage of these techniques is the fact that there was no intraoperative classification of the tumours into shamblin's groups. [19] Had this been done it could have prepared the surgeons to bypass or graft the carotid vessels in patients with Shamblin group II or III tumours. Another possible explanation for non-usage of these techniques is lack of a definite indication for their use as seen by surgeon intraoperatively.

OUTCOME OF SURGERY AND COMPLICATIONS.

Post-operatively and on subsequent follow-up 60% of the patients were presumed to have been cured since they did not exhibit any recurrence. Ten patients {33.3%} developed recurrence of tumour; of these one died. However, the percentage of patients with recurrence can drastically be reduced if the interventions suggested, i.e. preoperative embolisation of tumour vasculature, or radiotherapy, and carotid artery bypass grafting are undertaken. Fifty percent of all patients suffered hoarseness of voice post-operatively. This was followed by nineth and twelveth cranial nerve palsies (table 17). Vagus nerve palsy per-se accounted for only 10% . The cranial nerves, especially nineth, tenth and twelveth are closely related to the carotid sheath in the neck. Being a very vascular tumour CBT bleeds considerably at surgery, However with meticulous control of bleeding preferably by bypass grafting of the carotid vessels this complication should drastically be reduced. Interestingly there was no single patient with Horner's syndrome despite the close proximity of the sympathetic ganglion chain to the carotid body. Horner's syndrome is recognized as one of the complications of CBT surgery but is not universal and the fact that no patient was diagnosed to have it post-operatively may suggest that either it was overlooked or was not recognized during post-operative evaluation.

CONCLUSION:

1. Thirty patients (30) presented with neck masses which were diagnosed to be carotid body tumour (CBT) over an eleven year period, putting the annual prevalence of the condition at 2.72 patients.
2. In this study the female to male ratio of CBT occurrence was 7:3 ; this contrasts with the worldwide known ratio of 1:1; this however does not imply that CBT occurs more in females than in males in our setup.
   The age range of presentation, together with the peak age (45years) compares well with the documented world wide figures.
3. The majority of patients with CBT came from Central province and were, predominantly of Kikuyu origin. Apart from the fact that Central Province is close to Nairobi (and hence accessibilityto KNH is easier) and Kikuyus in particular are the most populous community in the country, no other factors were identified. Further studies need to be carried out to establish the existence of predisposing factors if any.
4. Altitude did not explain the development of CBT in our setup. Whether this factor is insignificant in the aetiology of CBT can only be confirmed by further studies.
5. Familial CBT was not established in this study; even the three patients with. bilateral CBT did not have a positive family history
6. All patients presented with a lump in the neck. Only one had experienced an inadvertent attempt at excision of the tumour in a peripheral facility.
   The fact that all these patients were referred to the main facility without attempts to excise the lumps elsewhere, except for the one case above is applaudable and should be encouraged .
7. Most patients presented to hospital relatively early before further advancement of the tumour.
8. All patients except one were adequately investigated to confirm the diagnosis; however the few patients (10%) who had plain x-rays done did not need them in the first place
9. In terms of operative management all modalities were not employed both preoperatively (radiation, embolisation) and intraoperatively (bypass, grafting) in order to maximize results.

RECOMMENDATIONS:

1) A Management protocol for all patients suspected to have CBT should be formulated. This will simplify data derivation in case of subsequent studies. Such protocol should include:-
    a) Patients identification and demographic data
    b) Patients' clinical data
    c) A standardized investigative approach - This will pre-empt the tendency to over-investigate a patient by e.g plan x-rays, ultrasound then angiography,and hence minimize unnecessary cost to the patient.
    d) A standardized management approach - This should include intraoperative categorization of patients into shamblin groups and hence a standardized operative approach to each group.
    e) A standardized post-operative and follow-up checklist. This will ensure that no complication is missed out.

2.) The successful management of this condition calls for a multidisciplinary interaction.With the presence of interventional radiologists in KNH for example, it should be possible to carry out pre-operative embolisation of tumour vasculature, which will improve the final outcome of the management of CBT.

3) For a hospital of KNH's magnitude it is unwise to operate on a patient without any investigations to back the diagnosis unless there is an exceptional reason to the contrary. This should be avoided especially in this era of increased patient education and litigation.

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APPENDICES:

APPENDIX - 1

DATA COLLECTION FORM (QUESTIONNAIRE)

CAROTID BODY TUMOUR AT KENYATTA NATIONAL HOSPITAL (1986-2000)

SERIAL NUMBER:

A. DEMOGRAPHIC DATA

(i) Name:.....

(ii) I/P number: .....

(iii) Age: .....

(iv) Gender: (Female = F, Male = M).....

(v) Tribe: .....

(vi) Nationality (Kenyan = K, Non Kenyan = NK) .....

(vii) Home District: .....

(viii) District of Residence

(ix) Altitude of District of Residence ..... feet above sea level

B. HOSPITAL DATES:

(i) Date first seen at KNH .....

(ii) Date of admission .....

(iii) Date of operation .....

(iv) Date of discharge/death .....

(v) Date if first follow up .....

(vi) Total No. of revisits after surgery .....

C. **HISTORY:**

(i) Presenting Complaint and Duration

Complaint	Yes = 1; No = 2	Duration
▪ Asymptomatic		
▪ Lump(s) in the neck		
▪ Neck pain		
▪ Neck discomfort		
▪ Hoarseness of voice		
▪ Dysphagia		
▪ Headache		
▪ Dizziness		
▪ Others (specify)		

(ii) Relevant past medical history Yes = 1; No = 2

- Previous neck surgery { }
- Previous neck irradiation ( )
- Previous treatment for cancer ( )
-

(iii) Family and social history

- No. of siblings ( )
- Ranking in family (1st, 2nd, 3rd etc) ( )
- Family Hx of neck mass ( )
- If yes to above: - brother ( )
  Sister ( )
  Son ( )
  Daughter ( )
  Parent ( )

Distant relative ( )

EXAMINATION FINDINGS (SIGNS)

(i) Local signs:

- Neck mass (Yes = 1; No = 2) ( )

If yes

- Site
- Size
- Shape
- Surface
- Solidity (consistency = soft, firm hard)
- Skin overlying it
- Mobility (vertical; lateral)
- Tenderness
- Temperature
- Pulsatility
- Bruit
- Bilateral

If bilateral characterise the other mass as above

(ii) Distant signs (Yes = 1, No = 2)

- Parapharyngeal space mass [ ]
- Horner's syndrome [ ]
- Cranial nerve palsy IX [ ]
  X [ ]
  XI [ ]
  XII [ ]

(ii) Systemic signs (specify)

- Gastrointestinal tract

- Respiratory system
- Cardiovascular system
- Central nervous system
- Musculoskeletal system

(iv) Functional evaluation done [ ]

D. RADIOLOGICAL ASSESSMENT:

Investigation Result

- Plain x-ray (skull and neck)
- Ultrasound
- Angiography
- CT Scan
- MRI

F. **DEFINITIVE MANAGEMENT** (Yes = 1, No = 2)

- Observation [ ]
- Radiotherapy [ ]
- Surgery [ ]
- Combination (specify) [ ]

(i) Surgical management

Pre-operative intervention: (Yes = 1, No = 2)

- Radiation
- Embolisation

Operative intervention (Yes = 1, No = 2)

- Partial excision [ ]
- Total excision [ ]
- Inoperable [ ]
- Carotid grafting done [ ]
- Carotid bypass done [ ]

Shamblin Group (I, II, III, not classified) [ ]

Histological results: - Carotid body tumour [ ]

(ii) Outcome of surgery (follow-up)

- Cured [ ]
- Recurred (Ipsilateral) [ ]
- Recurred (contralateral) [ ]
- Patient died [ ]

(iii) Complications

- Cranial nerve palsy - IX [ ]
  - X [ ]
  - XI [ ]
  - XII [[

- Horner's syndrome [ ]
- Hoarseness of voice [ ]
- Cerebrovascular accident [ ]
- Others (specify) [ ]

APPENDIX II

DUMMY TABLES AND CHARTS

Examples of tables and charts used in data presentation

Fig 1: Age distribution of patients with CBT:

AGE GOUP (YEARS)	NO. OF PATIENTS	% OF TOTAL
5-14		
15-24		
25-34		
35-44		
45-54		
55-64		
65-74		
75-		

Fig 2: Sex distribution of patients with CBT

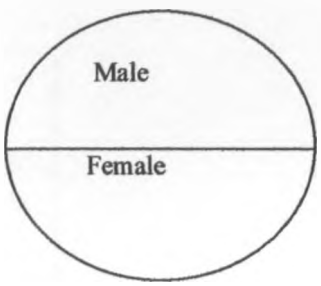


Figure 3 Symptoms of patients with CBT

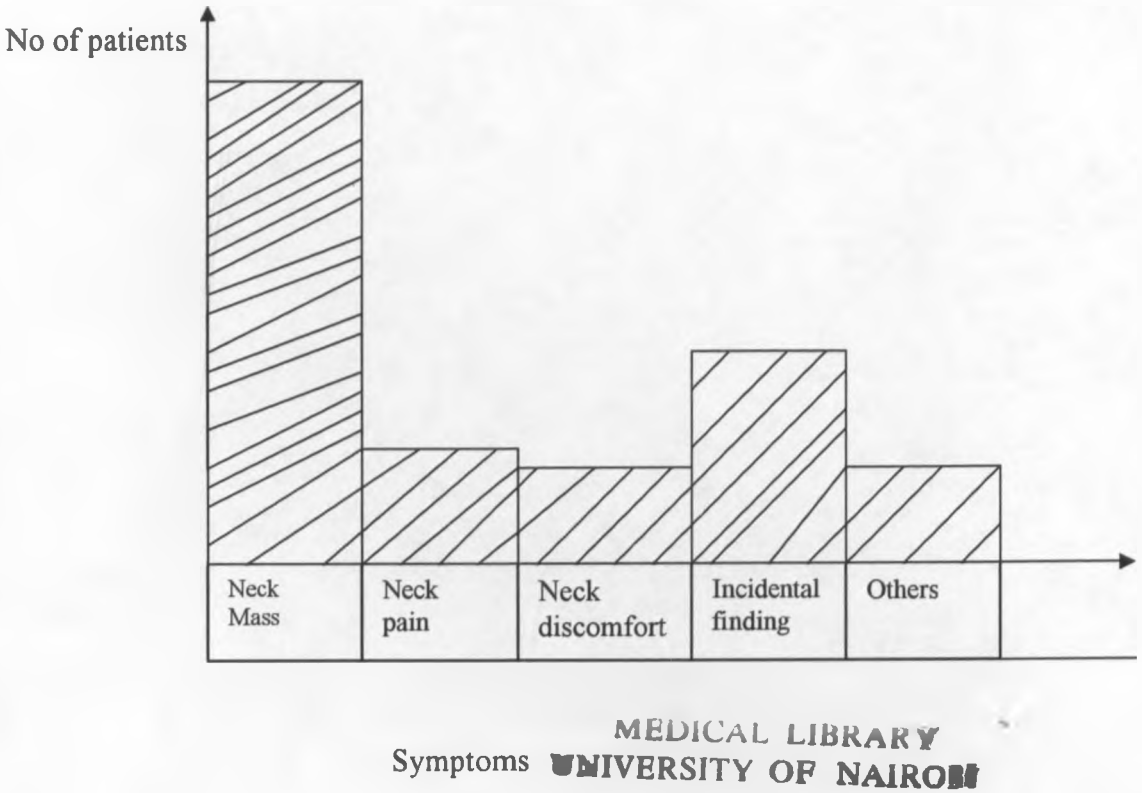
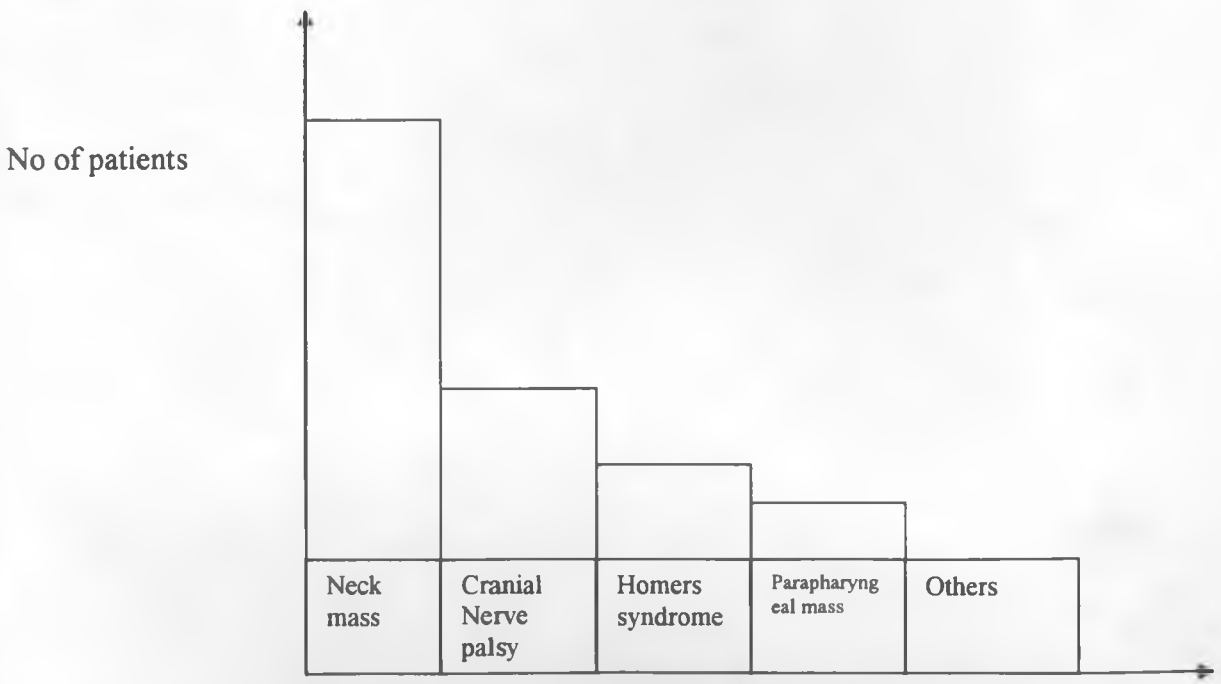


Fig 4: Signs of patients with CBT



Dr. Fred Welch Muleshe
Dept. of Surgery
Faculty of Medicine
University of Nairobi

Dear Dr. Muleshe,

RESEARCH PROPOSAL " A REVIEW OF THE MANAGEMENT OF CAROTID BODY TUMOUR AT KENYATTA NATIONAL HOSPITAL (JAN 1990 - DEC 2000)" (P52/6/2001)

This is to inform you that the Kenyatta National Hospital Ethical and Research Committee has reviewed and **approved** your above cited research proposal.

On behalf of the Committee I wish you fruitful research and look forward to receiving a summary of the research findings upon completion of the study.

This information will form part of data base that will be consulted in future when processing related research study so as to minimise chances of study duplication.

Thank you.

Yours faithfully,

PROF. A. N. GUANTAI
SECRETARY, KNH-ERC

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