



## Clinical Research

# Identifying Factors Influencing Decision Making in Patients Diagnosed with Carotid Body Tumors: An Exploratory Study

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**Background:** Carotid body tumors (CBTs) are rare highly vascularized and slow enlarging tumors arising from the paraganglionic tissue at the carotid bifurcation. Main treatment options for CBTs are surgical resection or “wait and scan” strategy. The choice for either strategy may be equally good medically in many patients. A structured “shared decision making” (SDM) might be helpful for guiding patients.

**Objectives:** To develop an SDM strategy for the surgical treatment, we aim to (1) identify considerations and factors involved in the decision making of patients with CBTs and (2) evaluate the current practice in our clinic and explore the opinions of patients on their treatment.

**Methods:** This exploratory study was conducted in patients of the Leiden University Medical Centre (LUMC), The Netherlands. Patients who met the inclusion criteria were invited for a semi-structured interview. All conversations were fully audiotaped and transcribed.

**Results:** Fifteen patients were included and interviewed. Ten of these patients underwent previously surgical resection of at least one tumor. Five patients underwent the wait and scan policy. The most important factors influencing decision making in CBT treatment are family, fears, co-consultants, and doctor-patient relationship.

**Conclusions:** This study has identified the factors influencing decision making in CBT and should be considered during consultations. The decision for surgery or not was mainly influenced by physician preferences and family members’ prior experiences.

## INTRODUCTION

Carotid body tumors (CBTs) are rare highly vascularized and slow enlarging tumors arising from paraganglionic tissue at the carotid bifurcation.<sup>1</sup> In 10–

50% of the cases, paragangliomas can be associated with mutations in the succinate dehydrogenase (SDH) complex subunits.<sup>2</sup> CBTs are frequently autosomal dominant and highly penetrant in case of a genetic defect; in some cases, it also arises sporadically.<sup>2</sup> CBT in an early stage is mostly asymptomatic and clinically silent, whereas in a later stage of development, they can cause dysphagia and deficits of the cranial nerves: facial nerve, glossopharyngeal nerve, vagus nerve, accessory nerve.<sup>3,4</sup> Overall CBTs are considered benign; however, when histological reviewed, 4.1% of the tumors were found to be malignant.<sup>5</sup> Metastasis occurs in 2% of the patients diagnosed with CBT.<sup>6</sup>

The tumor size and the degree of involvement of the internal carotid artery has been described by Shamblin et al. and is related to the incidence of

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postoperative complications.<sup>7,8</sup> The craniocaudal approach is the surgical technique of choice in our institution as it limits blood loss and facilitates safe CBT resection.<sup>9,10</sup> CBTs are slowly growing indolent tumors and can safely be followed.<sup>11</sup> In cases with a large or fast growing tumor wherein surgery is not an option because of concomitant disease or extensive local advancement, radiotherapy can be an option.<sup>12,13</sup>

Because the treatment options in patients with CBT are often equivocal, “shared decision making” (SDM) could lead to more satisfying decisions. The benefits and harms of each treatment option will be discussed where the patients’ preference and value will be taken into account.<sup>14</sup> Knowing which factors from patients’ perspective have an influence on the decision making is essential. Kunzel et al. re-evaluated the decision algorithm implemented by their German institution in the treatment of cervical paragangliomas.<sup>15</sup> However, this study did not include patients’ preferences or experiences. Previous studies have only included clinical factors in decision making.<sup>16</sup> Studies on shared decision making in several cancer types have revealed important factors influencing patients’ and physicians’ preferences that can benefit decision making during consultation.<sup>17,18</sup>

To develop a useful SDM strategy for the surgical treatment of patients with CBT, we aim to (1) identify considerations and factors involved in the decision making of patients with CBTs and (2) evaluate the current practice in our clinic and explore the opinions of patients on the clinical management decision strategy.

## METHODS

### Patient Selection

This exploratory study was conducted in patients of the Leiden University Medical Centre (LUMC), the Netherlands. Most patients with CBTs in the LUMC go through a specific clinical path for patients with paragangliomas of the neck. The selection for this study was taken from a historical database of patients with head and neck paragangliomas. In addition, surgical records for CBT resections were reviewed. Patients were invited to an interview if they met the following inclusion criteria: (1) Patients had a CBT and their check-up (consisting out of MRI-scan and consultation) is performed less than 1 year or a history of CBT resection that took place within 3 years of the interview; (2) Patients did not have a history of current or past paragangliomas other than CBTs; (3) Patients were

aged 18 years or older and resided in the Netherlands during the course of the study. Patients were excluded if they underwent therapies other than “wait and scan” or surgery at any time, such as radiotherapy or experimental procedures. Patients willing to participate were offered the option of choosing the location of the interview. The same investigator conducted all interviews. Interview by telephone was not chosen as an option as it limits direct interaction between the patient and the interviewer. Patients were sent a letter of invitation to participate in this study. The invitation was posted to their address of residence which was retrieved from the hospital records. Nonresponders were contacted by phone up to 2 times after 4 weeks.

### Wait and Scan

All patients with a CBT, referred to the department of vascular surgery, were given the option to either undergo surgery, or prolonged follow-up according to the “wait and scan” protocol and in some cases radiotherapy.

Once a month, a multidisciplinary meeting is scheduled to discuss the cases with the departments of vascular surgery, genetics, radiology, and otorhinolaryngology. Patients presenting at first consultation with relatively small CBTs (<2, 5–3 cm) are advised to wait and see the natural course of the tumor. Unless surgery is an explicit wish, monitoring tumor growth is the treatment of choice in small tumors. Annually or once every 2 years, an MRI scan of the head and neck area is performed to objectify growth of present tumors and detect additional paragangliomas. The MRI results are discussed during a consultation and a new decision is made; to continue with the policy “wait and scan” or convert to surgery. In case of new symptoms, progression of existing symptoms, or when there is strong wish to undergo surgical resection, surgical options are reconsidered. Surgical procedures are executed by or under direct supervision of at least one vascular surgeon with a special expertise in the field of head and neck surgery. As a tertiary referral center, 5 to 8 surgical resections are performed annually, by 2 dedicated surgeons. In Leiden University Medical Centre, resection of CBTs is performed by a modified dissection technique as described by vd Bogt et al. Resection starts in a craniocaudal fashion from skull base to carotid bifurcation. This modified dissection technique reduces the risk of postoperative morbidity and mortality.<sup>10</sup> Patient follow-up was performed by the Department of Otorhinolaryngology at the outpatient clinic.

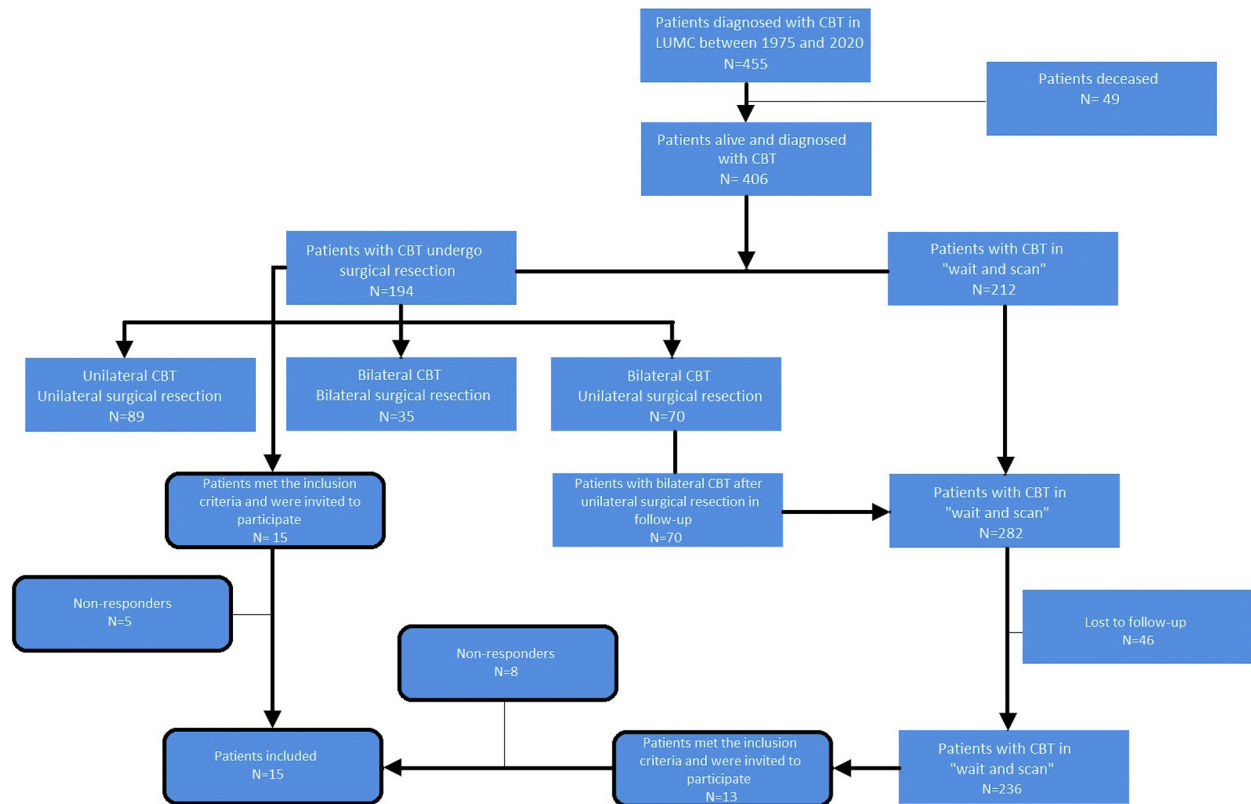


Fig. 1. Flowchart of patient inclusion.

### Data Collection and Content Analysis

Participants were interviewed in Dutch in a semi-structured fashion. Re-evaluation of the questions was done after the first 4 patients. Questions based on previously observed patterns in answers were added to the lists as a standard. The final version of the interview questions can be found in [Appendix A](#) (translated in English). All data were rendered completely anonymous in a retrospective database and patients were registered by a number from one to fifteen for study purposes. Audio tapes of interviews were transcribed verbatim and analyzed according the principals of "content analysis." Content analysis is a method for describing the content of communications in an objective and systematic manner.<sup>19,20</sup> Declaration of no objection has been obtained by the ethics committees of LUMC and this study was approved by the Medical Ethical Committee.

### Statistical Analysis

There is no statistical program used. Based on their distributions, continuous variables are presented as

mean  $\pm$  standard deviation or median. Dichotomous and categorical data are expressed as numbers and percentages.

## RESULTS

### Baseline Characteristics

Between 1975 and 2020, there were 455 patients diagnosed with CBT (Fig. 1). Forty-nine patients were deceased and excluded. Twenty-eight patients met the inclusion criteria and were invited to participate in the study. Fifteen patients underwent CBT resection and thirteen patients still had a CBT in situ and were followed in the wait-and-scan regime. Ten patients spontaneously responded to the invitation and were willing to participate. The remaining patients were contacted by phone twice after 4 weeks, which led to the inclusion of another 5 patients. Two patients declined the invitation and the remaining 11 patients could not be contacted. A total of 15 patients were eventually included and interviewed. Ten patients underwent previously surgical resection of at least one tumor (one patient had bilateral resection). There was a mean interval

of  $17 \pm 11$  months and a median of 16 months between the surgery and interview. Five patients were followed according to the wait and scan policy. Nine patients were female. The mean age at inclusion was  $43 \pm 19$  years. Eleven patients carried the SDHD mutation, 2 patients had no proven genetic predisposition and 2 patients had chosen not to undergo DNA testing.

## INTERVIEW RESULTS OF OPERATED PATIENTS

### Preoperative Assessment

All included patients ( $n = 10$ ) were seen at the outpatient vascular surgery clinic by the operating surgeon to discuss the treatment options. All but one patient had made the final decision to undergo the surgery during the consultation. One patient had already decided before the consultation took place. The main reason to undergo an operation as reported by patients was tumor growth ( $n = 7$ ). Three of the included patients had undergone resection of a nonprogressive tumor upon their own request. In 2 cases, this was due to a family history of metastasized CBTs. The final patient opted for surgery due to the symptoms of the tumor, among which mechanical obstruction during swallowing and, as she stated herself, chronic fatigue.

### Information and Accessibility

Patient education was performed verbally, illustrated with MR images in all cases during the consultation. One patient reported the current way of patients' education to be sufficient. However, 8 patients reported the absence of proper information material. All patients except for one had not received folders to bring home and reflect on. Five patients however report the need of tangible material that is applicable to their case to bring home and reflect on. Three patients stated that information online has, in their opinion, proven to be too overwhelming and complex.

Treatment options such as surgery and wait and scan are mainly elaborated during the consultation. Radiotherapy is not offered as a standard treatment for CBT in the LUMC; however, 7 patients were preoperatively informed about radiotherapy as treatment option by the otorhinolaryngologist. In five of 7 cases, exploring radiotherapy as treating option was initiated by the patient. Only 2 patients had been referred to the radiotherapist for a consult on radiotherapy in paragangliomas. However, upon inquiry, 7 patients reported the wish for information

on radiotherapy by the physician to be standard during consultation because radiotherapy is readily reported as a treatment option on the Internet.

Four patients felt that information on experimental procedures (when present) should be given as a standard as well. The other 6 patients however felt that too many options would be too confusing and that experimental procedures should only be discussed if it was an actual treatment option for them.

### Evaluation of Preoperative Consultation and Decision Making

All patients reported that they felt at ease with their physicians and could ask all their questions without restriction. Doctors were described to be accessible, clear, elaborate, and trustworthy. Patients agreed on the fact that the most important quality of their doctors was their reliability. "If I would not have been able to see that he was confident that the surgery would be a success, I would not have agreed with the procedure" was a common statement. All patients stated that pros and cons, including risks, of the procedure were discussed elaborately before deciding to operate. Four of the surgical patients stated to have considered an operation earlier on if they had known how they are today. One patient however criticized that although the risks of an operation were discussed elaborately, the risks of waiting were not discussed at all. He stated that if he had known that with progression of the tumor, that is, a larger tumor, the operation risks increased, he would have had the surgery earlier. Another patient stated that she would have had the operation earlier, if she had the chance. She stated that she wanted the surgery a few years before the actual procedure, but the physician did not take this option into consideration. When inquired, she stated that her physician made decisions before the operation solely and it had not been a shared decision.

### Operative Outcome, Hospitalization, and Recovery

The mean time interval between the first observation of the CBT and the operation was 3.8 years (range 0 to 10 years). In total, 11 CBTs were removed in 10 patients. One patient had bilateral CBT and underwent a resection of the CBT on both sides with an interval of 3 months. Most of the resected tumors were Shamblin type II tumors, 2 were Shamblin type I, and only one tumor was Shamblin type III. Transient cranial nerve (CN) damage was present in 2 patients. In one of the patients, accessory nerve, and in the other patient, it

was the marginal ramus of the facial nerve. These dysfunctions resolved spontaneously within 6 months. One patient had permanent failure of the vagal nerve due to the concurrent resection of a glomus vagal tumor (GVT). Two patients suffered from first-bite-pain syndrome. One of these patients recovered spontaneously within 3 months; in the other patient, it is still present one year after surgery. None of the patients suffered from postoperative stroke. Postoperative hospital admittance ranged from 2 to 5 days. Most patients resumed their work after a sick leave of 3 weeks. Twenty percent resumed their normal rhythm after an absence of 3 months. Three patients however had a leave of one year.

## INTERVIEW RESULTS OF “WAIT-AND-SCAN” PATIENTS

### Patient Assessment

Five patients with 8 tumors were treated according to the “wait and scan” policy (3 patients had bilateral CBTs). Follow-up included three type I, three type II, and two type III tumors according to the Shamblyn classification. The interviewed patients had an average follow-up of 4.4 years and median of 3 years (range 1 to 11 years) and their last check was performed for less than 1 year from the time of interview with a mean of  $7 \pm 4$  months and median of 10 months. Their last check-up consists out of an MRI scan and a consultation. In the consultation, the results are evaluated and a (new) decision is made. None of the tumors were progressive over time. Two patients reported to have symptoms. One patient had mechanical objections without limitations in her daily routine. Another patient stated to feel more tired and experienced problems with concentrating ever since the tumor had been observed. Catecholamine excess was not present in any of these patients.

### Treatment Options, Information, and Accessibility

Patients were followed with annual MRI scans on which potential progress of the tumor was evaluated. Patients were informed of the results either at the outpatient clinic in person or per telephone consultation, depending on their residential distance. All patients had consulted a vascular surgeon to elaborate on the option of surgery at least once in the last 4 years. They all stated that pros and cons and possible risks of surgical treatment were

discussed clearly and that there was enough space for rebuttal.

One patient was informed about treatment with radiotherapy by the otorhinolaryngologist. The other patients were only offered the option of surgery versus “wait and scan”—Three did however regret this and stated that all treatment options should have been discussed clearly. Similar to the surgical patients, however, they felt that too many options would be overwhelming and that experimental procedures should only be included when applicable to that specific patient.

### Decision Making

The most common reason for patients to postpone surgery was the absence of growth of the tumor. They stated “you could live up to a 100 without ever needing surgery.” “Why operate and perhaps cause unnecessary damage?” was a common statement. Another common idea was that in the future, more advanced surgical procedures will be developed that have lower risks of CN damage and stroke. One patient was reluctant to be operated on due to the poor operative outcome of a sibling. This was however not his main reason, which was in fact the absence of growth. Although most patients stated that the decision not to operate was a mutual decision by physician and patient upon information provided by the vascular surgeon, 2 patients stated that the decision was not mutual. They stated that their physician solely decided not to operate. Both patients felt that they did not have a say in the decision and felt that surgery might be a good option in their case. They noted that the option of an operation had since their first consult not been discussed with them anymore. For one of the patients, this was 10 years ago. They felt the need for the physician to bring up the subject of the treatment option every once in a while, as they did not feel the space to do so themselves.

## DISCUSSION

The primary aim of this study was to identify the factors, from a patient’s point of view, which have an influence on their decision making regarding a treatment for CBT. The main factors influencing their decision as described in this study were family, fears, co-consultants, and doctor-patient relationship apart from “medical” factors such as growth of the tumor. To our knowledge, our study is the first study examining factors influencing decision making in CBTs in patients.

Patient anxiety and fear about postoperative complications affect their choice about having an operation despite comprehensive preoperative counseling.

On the other hand, the experience and outcome of their relatives affects the patients' choices for treatment. Although every CBT has to be assessed individually, the poor outcomes of their family are seen as equal possible outcome for themselves.

Most patients had consulted with their partner or other family members before making their final decision for therapy. Patients frequently spontaneously stated that they had high confidence in their doctors. This confidence underlines the need for a good doctor-patient relationship and confidence in the treating physician, as stated by Hofstede et al.<sup>21</sup>

The secondary aim of this study is to evaluate the current practice in our clinic and explore the opinions of patients on the clinical management decision strategy. From a medical point of view, watchful waiting and surgical resection are equal in benefits and harms in CBT of stable size. This study confirmed again that patients need well-tailored written information they could reflect on at home. Providing adequate information was defined as a critical factor in successful shared decision making by Hofstede et al.<sup>21</sup>

Elaborate information was provided by the doctor on the disease, the natural course, and the treatment options. In only one out of all 15 cases, an information brochure was handed out to read at home. All other patients searched for additional information about CBT on the Internet. They had a hard time tailoring the information (found on the Internet) to their own situation.

Shared decision making is a choice of strategy where the physician and patient make decisions jointly, weighing the best available evidence regarding different treatment option.<sup>17</sup> Most of the patients in this study felt that they had decided on their treatment in consultation with their physician; however, the patients that disagreed on this point stated that they needed more control in their disease and choice of treatment.

One of the most striking findings was that patients reported to be interested in radiotherapy. Although radiotherapy is not a conventional treatment option for CBTs, it is increasingly reported in literature. Patients nevertheless wished to receive information on radiotherapy, seeing as it is a possibility. Patient education is therefore important and should take place. Patients should be informed about radiotherapy and informed according to their personal situation so doubts and questions can be detected and dealt with.

Information provision and a good relationship are the most important conditions for SDM perceived by both patients and professionals. If the physicians know what factors have an influence on the decision making, these factors can each be elaborated. Such a patient-centered outcome benefits patient satisfaction and facilitates a sustainable doctor-patient relationship.

Apart from the factors of decision making, we identified 2 postoperative morbidities that patients encountered but were not sufficiently informed about preoperatively. This concerns fatigue and the First Bite Syndrome. A recent study by Van Hulsteijn et al. indicated a significantly impaired quality of life (QoL) among patients with paraganglioma.<sup>22</sup> Their study concluded that, compared to controls, patients reported a significantly impaired QoL, mainly on fatigue and physical condition subscales. This was especially the case in patients with CBTs.<sup>22</sup>

In addition, we have encountered 2 patients with first bite syndrome (FBS). Recent reviews describe FBS sporadically as a complication of CBT surgery.<sup>23,24</sup> These are however small series, but FBS is an outcome that patient wants to be informed about preoperatively.

### Limitations

In this study, the number of included patients was low. However, considering the rareness of this tumor and our inclusion criteria, the results are representative for this population.

The interviews took place several months after their surgery or last control at the hospital. This study can therefore be limited by a recall bias, which is tried to limit by only including patients that had a surgical resection within 3 years from time of interview.

There are no readily validated semi-structured interview questionnaires to meet the goals of this study. Patients have however proven to give similar answers. Comparable answers were a result of using thematic content analysis, indicating that collected data were valid.

### CONCLUSION

This study has identified fear, family influence, and co-consultation as major factors influencing decision making in CBT. Patients' decision is influenced by physicians' preferences and family members' prior experiences. These factors can be taken into account during consultations and SDM. There is also a difference in how patients feel about their involvement in decision making and how

physicians think that they are involving patient to make a decision in their treatment. To provide a well-tailored information for the patient to review at home, we have developed an information pamphlet that covers all the basic information about CBT and treatment options.

## SUPPLEMENTARY DATA

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.avsg.2020.05.044>.

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