Information for patients, family and carers: Skull base cancers

What is a skull base cancer?

Your doctor will confirm with you the nature of your skull base cancer. Many ‘skull base cancers’ are actually malignant tumours that start in either the linings of the nose/air sinus cavities, the ear area, structures of the eye, or facial skin or scalp, and then grow to involve the base of skull. Sometimes the skull base cancer may have spread from somewhere else, i.e., a metastasis from a ‘primary’ cancer elsewhere, e.g., from a lung, kidney, breast, or skin cancer. There are many different types of skull base cancer and many are rare. The exact nature of your cancer will need to be confirmed before a treatment plan can be discussed.

What is meant by skull base?

The base of the skull is that part of the skull that runs from your eyes to the back of your head. It separates (and supports) the brain above from the eyes, nose, ears, and neck below. Important blood vessels such as the carotid arteries supplying the brain and important nerves including ‘cranial nerves’ pass through the skull base. The cranial nerves deal with functions including smell/taste, vision, sensation of the face, facial muscle movement, hearing/balance, swallow, and voice. The base of skull is closely related to important parts of the brain above and is also closely associated with your eyes, nose, mouth cavity, and ears below.

What causes skull base cancers?

Many cancers involving the base of skull are very rare and the exact cause is not known. A proportion of cancers that start in the mouth/nose cavities known as 'squamous cell carcinomas' are considered smoking-related. Occasionally, a skull base cancer can be due to an occupational exposure to a known hazard, e.g., wood dusts for joiners/wood carvers. Rarely, there is a genetic cause or there may have been radiotherapy to the area concerned many years previously.

What symptoms may occur?

Initial symptoms generally reflect the structure the cancer started off in. For example, a cancer starting off in the nasal cavity might manifest as a persistently blocked nose over weeks with nasal discharge, bleeding down nose followed by increasing facial pain, facial swelling, and eventually bulging of the eye. A cancer of the ear canal might manifest as an ear discharge with
bleeding, followed by increasing ear pain and even muscle weakness on that side of the face. A skull base cancer may also cause increasing one-sided facial pain and development of facial numbness. Unfortunately for a variety of reasons, patients with skull base cancers often have symptoms persisting for many months before coming to our attention.

**How is a skull base cancer diagnosed?**

A skull base cancer is diagnosed by performing scans of the head and neck area and than a biopsy. Usually the head and neck scans performed are both CT, which gives the best information about bone, and an MRI, which gives the best information about soft tissues. Additional imaging will also be performed both to confirm that your cancer has not spread from somewhere else (a metastasis) and also to check whether there has been spread elsewhere in the body. Such additional imaging is usually a body CT and possibly a PET scan and/or bone scan. Only once all these tests have been performed will your doctors be able to tell you exactly the nature of your skull base cancer and the type of treatment that will be required.

**What are my treatment options?**

Some skull base cancers are best treated with skull base surgery followed by radiotherapy. Other skull base cancers may also require chemotherapy. Occasionally radiosurgery or proton therapy is also a possibility. For some skull base cancers, skull base surgery may not be appropriate or required. Your doctors will outline a proposed treatment plan once they have confirmed your diagnosis.

Treatment objectives can differ depending on your cancer and will be made clear by your treating doctor. Sometimes a treatment will be offered on the basis that it is done with ‘curative intent’. Sometimes curing the cancer is not a possibility but treatment will delay the progression and effects of the cancer. In particular, treating to achieve ‘local disease control’, even if the cancer may become a problem elsewhere. Trying to get local disease control can be important as a progressing untreated skull base cancer can result in severe pain, unpleasant ulceration of face/head, and other difficult issues that very much subsequently impact on quality of life.

**Surgery**

Major skull base surgery to remove a skull base cancer is complex and demanding. Your doctor will indicate whether major skull base surgery is an option and the specific issues involved.

Such surgery is usually lengthy lasting many hours, may be disfiguring (e.g., loss of eye or ear, facial asymmetry), and may result in significant loss of function (e.g., loss of smell/taste, visual problems affecting an eye, swallow
difficulty). If the surgical team can minimise any cuts on the face by using a keyhole or minimally invasive ‘endoscope-assisted’ technique, they will.

The surgery is not just about removing the cancer. The surgeon has to also close the hole made in the skull base as a result of the cancer removal. Sometimes muscle or other tissue is taken from somewhere else known as the ‘donor site’ (e.g., back, arm, abdominal wall) to close the hole with the muscle/tissue blood supply plumbed into blood vessels in the neck. This type of repair is called a ‘free flap’.

Sometimes a ‘neck dissection’ is performed, i.e., removal of lymph glands in the neck, in anticipation of where your cancer may spread next. Whether this is done depends on the type of your cancer and the presence of any enlarged lymph glands on your scans.

You may have a temporary ‘tracheostomy’ placed at the time of your surgery, i.e., where a breathing tube is inserted into your wind pipe at the base of your neck. This is done where there are worries about chest problems after surgery or where the surgery will likely result in swelling around your upper airway.

A temporary feeding tube known as a ‘percutaneous enterogastrostomy’ or ‘PEG’ for short may be inserted prior to your major skull base surgery or radiotherapy if your doctor thinks that there will be temporary difficulties with swallowing as a side effect of treatment.

Problems that need to be addressed after major skull base surgery are common. Three to four out of ten patients having major skull base surgery will have such problems. Potential problems include clot in the leg/clot in the lung (‘DVT/PE’) 1:100, infection 1:100, stroke/blood clot (with the possibility of permanent paralysis and also having a small risk to life) 1:100, brain swelling 1:100, epilepsy 2-3:100, skull base reconstruction/free flap failure 1:10, brain fluid leakage/meningitis 1:20, ‘donor site’ problems 1:10, and chest infection 1:10. There are a whole range of measures that the surgical team take to stop problems happening and a whole range of measures that are taken to reduce the impact when problems do happen. You will spend an initial period in the neuroscience intensive care/high dependency unit after your surgery (see patient information leaflet on Neuro-Intensive Care Unit).

We would expect you to be in hospital for about three to four weeks after your surgery. Problems after surgery will extend your hospital stay beyond this period.

Your doctor may offer additional ‘reconstructive’ options once your cancer treatment, including any radiotherapy, is completed. Such options are discussed if there are significant facial appearance issues, eye problems, or facial muscle paralysis.
Radiation therapy

Radiotherapy, similar to that used in cancers elsewhere in the body, is often used for skull base cancers. A specific technique ‘intensity modulated radiotherapy’ or IMRT is used to focus the radiation on the tumour. IMRT may be given before or after major surgery or alone without surgery as a treatment for your cancer. Your doctor will clarify the extent of any radiotherapy in your treatment plan.

The radiotherapy treatment will be supervised by a specialist doctor known as an oncologist. Unless you have a treatment centre closer to home, the radiotherapy treatment would take place at Weston Park hospital in Sheffield. Treatment begins with the making up of a ‘mask’ so as to accurately deliver the radiotherapy. Treatment is daily over a number of weeks.

If you are having major skull base surgery followed by radiotherapy, your doctor will be aiming to start radiotherapy from six weeks after surgery. If radiotherapy is carried out too early, it can cause wound healing problems. However, radiotherapy does have to be carried out relatively early to ensure the greatest benefit in control of your cancer.

Side effects are common and include skin irritation/redness, swallow difficulty, blurring of vision, dryness of eye (s), and fatigue.

Stereotactic Radiosurgery

Stereotactic Radiosurgery (STRS) is a radiation based treatment and is sometimes used for skull base cancers in specific circumstances. Your doctor will indicate whether STRS is an option.

STRS involves the fixation of a stereotactic metal frame to the skull usually under local anaesthetic, repeating an MRI scan, and then treating the tumour using highly focused beams of gamma radiation.

Stereotactic radiosurgery is generally a single treatment planned and delivered all in one day with a one to two night hospital stay. This is carried out at the National Centre for Stereotactic Radiosurgery in Sheffield (see www.gammaknife.org.uk or patient information leaflet on Radiosurgery).

Chemotherapy

For some skull base cancers your doctor may recommend chemotherapy, i.e., cancer treatment with drugs. The type of chemotherapy and when/how much is given depends on the type of cancer. Side effects of chemotherapy are common and include feeling sick, loss of appetite, fatigue, and increased risk of infections with any temporary dropping of blood counts.
Proton therapy

Proton therapy is a very specialised type of radiation treatment used for a small number of rare skull base cancers, specifically chordoma and childhood rhabdomyosarcoma. Patients who are suitable for this type of treatment are sent abroad as there is no treatment facility currently in the UK. Further information can be found at: http://www.specialisedservices.nhs.uk/serv/proton-beam-therapy.

What should my expectations be over the long term?

Your doctor will make it clear to you if the treatment plan being offered is for local disease control only or is attempted curative. If your proposed treatment plan is attempted curative, generally a combination of skull base resective surgery and radiotherapy, then we would anticipate that half of patients undergoing such treatment will be alive at five years. For most cancers, achieving the five year milestone is considered a 'cure'. One exception is a type of cancer called adenoid cystic carcinoma that can return beyond five years after initial treatment.

If your cancer does recur, there may be some additional options such as chemotherapy or radiosurgery. However, achieving a 'cure' becomes unlikely in such circumstances.

Can I drive with a skull base cancer?

If you have been diagnosed with a skull base cancer, you will not be allowed to drive for a period of time if you have had an epileptic fit, have significant loss of vision affecting both eyes, weakness in the arms or legs, episodes of confusion, or have had recent major surgery.

You do need to contact the DVLA as early as possible after you have found out about your skull base cancer.

If you have contacted the DVLA previously and they have permitted you to drive, you will need to contact them again if your condition with respect to your skull base cancer changes, e.g., you have an epileptic fit, undergo further major surgery, or have disease recurrence.

The sort of period that you may not be allowed to for drive is six or twelve months. It is the DVLA that makes the decision concerning your driving including for how long you are not allowed to drive, NOT your consultant neurosurgeon.

The hospital will not contact the DVLA. It is your legal responsibility to do so. You can contact the DVLA by phone on 0300 7906806 or at www.dvla.gov.uk. You should also contact your motor insurance company.
Is there anybody I can talk with to get further advice and information?

You will be assigned a skull base ‘key worker’ who can provide further advice and information. Your assigned key worker is usually either your consultant neurosurgeon and/or skull base specialist nurse. They should provide you with a means of contacting them. They will also provide you with copies of correspondence such as your clinic letters for your own records and additional information material such as further patient leaflets referred to within the text above.

If you have been referred from a Head and Neck Cancer or Sarcoma service or are followed up subsequently by a Head & Neck Cancer or Sarcoma service, you will also have a Head and Neck Cancer or Sarcoma key worker, usually a clinical nurse specialist, associated with that service. For example, patients from Sheffield can contact the Macmillan Head and Neck Cancer nurse specialist on 0114 2268776 (see http://www.sth.nhs.uk/services/a-z-of-services?id=227&page=71) or the Sarcoma specialist nurse on 0114 2713478 (see http://www.sth.nhs.uk/services/a-z-of-services?id=187).

There is a Sheffield Cancer Information and Support Centre located on the Weston Park Hospital campus, open Monday to Friday 9.00 am - 4.00 pm, for information, advice, and support (drop-in/no appointment is necessary). You can contact them on 0114 226 5666 or visit their website www.cancersupportcentre.co.uk.

You may wish to discuss your diagnosis with your GP.

The hospital also provides a chaplaincy service for different faiths or indeed if you do not belong to any faith group. Note that if you are an inpatient the chaplains will not automatically visit you, even if you belong to a particular faith group. If you would like to see a chaplain, please ask a nurse, relative or friend to leave a message on 0114 271 4999.
In addition, there are some local, national and international patient information groups/charities who can offer further advice and information including:

- **Sheffield group for head & neck cancer: ‘Heads Together’**
  Tel: 0114 2261436

- **Sheffield group for sarcoma: Sheffield Sarcoma Support**
  Website: [www.sheffieldsarcomasupport.org.uk](http://www.sheffieldsarcomasupport.org.uk)
  Tel: 0114 2261436

- **Macmillan**
  Website: [www.macmillan.org.uk](http://www.macmillan.org.uk)
  Tel: 0808 8080000

- **Cancer Research UK**
  Website: [www.cancerhelp.cancerresearchuk.org/type/head-and-neck-cancer/](http://www.cancerhelp.cancerresearchuk.org/type/head-and-neck-cancer/)
  Tel: 0808 8084040

- **Adenoid Cystic Carcinoma Organization International**
  Website: [www.accoi.org](http://www.accoi.org)
  Tel: 001 888 2237983 (USA)

- **Adenoid Cystic Carcinoma Research Foundation**
  Website: [www.accrf.org](http://www.accrf.org)
  Tel: 001 781 2489699 (USA)

- **Sarcoma UK**
  Website: [www.sarcoma.org.uk](http://www.sarcoma.org.uk)
  Tel: 0207 2508271

- **Mouth Cancer Foundation**
  Website: [www.mouthcancerfoundation.org](http://www.mouthcancerfoundation.org)
  Tel: 01924 950950

- **Changing Faces**
  Website: [www.changingfaces.org.uk](http://www.changingfaces.org.uk)
  Tel: 0207 391 9270

*This information sheet is to be used only in the context of attendance at or admission to the Department of Neurosurgery, Royal Hallamshire Hospital, Sheffield Teaching Hospitals NHS Foundation Trust. No responsibility is held for the advice provided by external support groups listed or the information content provided on their websites. If in doubt, ask your doctor.*

Compiled by Mr Thomas Carroll, consultant neurosurgeon, 21st September 2012