After Retinoblastoma …..

Every year in the UK around 40 children are diagnosed with retinoblastoma. Increasing public and professional awareness and advances in treatment together with a centralised approach to care has resulted in excellent long term outcomes and over 95% of patients can expect to be cured of the condition. As a result, there is a growing population of over 2000 survivors of retinoblastoma in the UK, whose care and support, along with other survivors of childhood cancer, has become a priority area within the NHS through the government's cancer reform strategy. This article will highlight the purpose of follow up and what patients and families can expect from their long term follow up team.

On finishing treatment

On completion of treatment children with retinoblastoma will initially be followed up by their RB multidisciplinary team. At the end of treatment, feelings of relief and pleasure can be mixed with uncertainty and worry and a **detailed summary of the treatment** received with a clear plan of the care to expect over the coming months and years can help lessen some of these worries. The **care plan** will outline the purpose of follow up appointments, how frequently they will occur and who you can expect to see. If you are not offered an **end-of-treatment summary** and **care plan** then ask your team to provide one.

Immediately after treatment has finished the focus of follow up is:

- To check for retinoblastoma coming back in previously treated areas within the eye
- To check for signs of new tumours developing
- To ensure that there is no evidence of retinoblastoma having spread outside of the eye

During these early months off treatment you will have access to your **RB keyworker** who should be available to answer queries and provide continuing support and guidance.

In the longer term

With time, the risk of the tumour recurring either within the eye or elsewhere in the body falls and there is a shift in the focus of long term follow up with the emphasis moving towards monitoring for the late effects of treatment (chemotherapy and/or radiotherapy) and ensuring good health in the future, particularly in those patients with heritable RB. During this period, your care is likely to transfer from the RB team, in London or Birmingham, to a Late Effects team who have expertise in long term care and who are likely to be based in your local paediatric oncology centre. This may happen from as early as one year from finishing

treatment up to 16 years, depending upon local arrangements. The focus of long term follow up is:

- **Monitoring for the side effects of chemotherapy**
  The chemotherapy drugs used to treat retinoblastoma have a range of side effects, some of which may occur many years after finishing treatment. These should have been discussed with you at the start of treatment. There are national recommendations for investigating these long term side effects which include:
  - Carboplatin: possible effect on kidney function and hearing
  - Etoposide :secondary leukaemia which is a rare occurrence but generally within 5 years of completing treatment
  - Vincristine: possible nerve damage

- **Monitoring for the side effects of radiotherapy**
  Radiotherapy has a number of potentially challenging long term side effects because of which newer treatments are being developed in order to try and limit its use. However, it is an effective treatment for RB and was the mainstay of treatment for many years and a significant number of patients have retained their eyes and vision thanks to its use. Long term complications can be cosmetic, hormonal or related to second cancers and the focus of follow up is to identify and treat these promptly.

- **Lifestyle advice and health promotion**
  One of the priorities in long term follow up care is to encourage and support young people to be aware of their own health risks and to minimise these through a healthy approach to life. Patients who are known to have the heritable form of retinoblastoma carry an alteration in their RB gene which predisposes them to other cancers later in life, this is not the case for patients with non-heritable or non-genetic RB.

  Although this can be a difficult and worrying discussion for many young people it is important to remember that our current knowledge is based upon patients treated many years ago and we hope that through patient education, alteration to treatment and healthy lifestyle choices it may be a very different story in another 40 years. A recent paper published in the British Journal of Cancer studied second cancers in patients treated in Britain since 1951. This paper confirms the risk for patients with heritable RB and identifies certain cancers which occur most frequently. These are bone and soft tissue tumours, melanoma of the skin and cancers of the bladder, uterus and lung. Many of these cancers are associated with the damaging effects of UV-light or tobacco. By raising awareness in young people, encouraging them to avoid smoking and limit their exposure to UV-light by careful use of high SPF sunscreen these risks could be significantly reduced in the future. For this reason, patients with heritable RB should remain under the supervision of a Late Effects team lifelong. This does not always mean regular hospital appointment but patients should have access to a Late Effects keyworker, know how to contact the late effects team if needed and understand the risks in order to make their own lifestyle choices.

On reaching adulthood

As young people achieve adulthood their care is likely to move to a Late Effects team within adult services. The process of transferring care from paediatric to adult services is called transition and will be offered to all patients who need it. The need for on-going care into adult life is tailored to each individual patient depending on their genetic status and the treatment they received.

Typical Long term follow up Outcomes following differing treatments for RB

Surgery Alone and Non-heritable RB:

- Follow-up during childhood
- Discharge at 16 – 18 years to GP care and self-management.
- Access to late effects team if required in the future

Chemotherapy and Non-heritable RB:

- Follow-up during childhood
- Transfer to adult late effects team at 16 – 18 years
- Eventual discharge to GP led care and self-management

Heritable RB or treated with Radiotherapy:

- Follow up during childhood
- Transfer to adult late effects team at 16 – 18years
- Remain under supervision with access to an adult late effects team lifelong

Summary

There has been a welcome shift in the provision of long term follow up care for retinoblastoma and providers have the challenge of ensuring that each survivor receives high quality, individualised Aftercare which will enable them to achieve their full potential in life. Change takes both time and resource and the speed and extent to which this is achieved will depend upon time, resource and both professional knowledge and public empowerment.

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