Primary Enucleation for Group D Retinoblastoma in the Era of Conservative Systemic and Targeted Chemotherapy: The Price of Retaining an Eye

Ido Didi Fabian¹,², Andrew Stacey¹,², Kenneth Johnson², Tanzina Chowdhury²,³, Catriona Duncan²,³, M. Ashwin Reddy¹,², Mandeep Sagoo¹,²

¹Moorfields Eye Hospital, London, UK, ²Retinoblastoma Service, Royal London Hospital, London, UK, ³Paediatric Oncology Department, Great Ormond Street Hospital, London, UK

Purpose: In this study we have quantified disease burden in terms of examinations under anaesthesia (EUAs) and compared the number of EUAs after primary enucleation versus conservative chemotherapy in group D retinoblastoma patients.

Methods: A retrospective analysis of 92 group D patients (104 eyes), of which 40 (40 eyes) underwent primary enucleation and 52 (64 eyes) were treated initially with systemic chemotherapy. Number, frequency and overall period of EUAs were compared using univariate and multivariate analysis.

Results: Primarily enucleated patients had an average 3-fold fewer (whole cohort) or 2.3-fold fewer (unilateral retinoblastoma) EUAs compared to the chemotherapy group (P<0.001). Primary chemotherapy, young presentation age, bilateral disease, multifocal tumours, retinoblastoma family history and germline cases were found on univariate analysis to correlate with increased number of EUAs (P≤0.019). On multivariate analysis, however, only primary treatment type and age of presentation were found significant (P≤0.001). Primarily enucleated patients also had EUAs less frequently (x1.4; P=0.022) and for a shorter period (x1.5; P=0.001). Of the 55 unilateral-presenting patients, a new tumour developed in the fellow eye only in a single (2%) case with known family history. The median follow-up time was 61 months (mean: 66, range: 14-156), in which time no cases of metastatic spread or death were recorded.

Conclusions: Group D patients’ families should be counselled regarding the significant 3-fold difference in number of EUAs following primary enucleation versus systemic chemotherapy when deciding on a treatment strategy. In this regard, primary enucleation would be most beneficial for older patients with unilateral disease.