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‘Threat to Vision Score (TTV Score)’ in children with Optic Pathway Gliomas (OPGs): UK experience from prospective Low Grade Glioma (LGG2) trial

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Introduction

OPGs arise in 15-20% of patients with neurofibromatosis type 1 (NF 1) while the term “sporadic” is reserved for those tumours presenting in non NF1 children[1,2]. 50-75% of children with NF 1 are asymptomatic at the time of diagnosis and symptoms, when present, vary depending on tumour location[3].

Visual loss is the most worrisome complication of OPGs[4]. Neuro-ophthalmic examination is a key component in the diagnosis and management of these tumours. Visual acuity (VA) has been recommended as the main functional outcome measure for NF1-OPG clinical trials[5]. Chemotherapy has become the favored initial treatment modality.

A TTV score was developed using UK data from LGG2 trial in order to establish whether this can predict final VA and drive management of OPGs.

Materials & Methods

114 paediatric patients with OPGs were identified after a search in national Low Grade Glioma database (LGG-UK). Subjects were identified from 21 participating centers in the UK.

VA was assessed by Teller Acuity cards, HOTV and LogMAR optotypes depending on age and cognitive ability. VA was considered normal if BCVA was equal or better to 0.2 LogMAR, while severe VA loss was documented if best corrected VA (BCVA) was 1.0 LogMAR or worse.

Neurofibromatosis status, location of tumour as per Dodge classification, best VA at presentation and age were considered the factors to take in account in creating a score ranging from 10 (minimum score) to 40 (maximum score).

Each component was weighted to reflect increasing severity (Table 1). The higher the score the greater the TTV.

Table 1

NF status (Weighted 4th important) Multiply score by 1		**Location of tumour** (Weighted this most important) Multiply score by 2		**Best visual acuity at presentation** (Weighted 2nd most important) Multiply score by 3		**Age** (Weighted most important) Multiply score by 4	
Non-NF1	2	Dodge III	3	<1.0	4	<1	5
NF1	1	Dodge II	2	0.8 - 0.6	3	1 - 2 yrs	4
		Dodge I	1	0.5 - 0.3	2	3 - 5 yrs	3
				0.2 or better	1	6 - 8 yrs	2
						8 yrs +	1

Results

All patients who scored > 30 underwent treatment with chemotherapy within approximately 2 months after diagnosis, while only 30% of subjects who scored < 20 received chemotherapy. The median time from diagnosis to initiation of treatment in these patients was 1.6 years (Table 2). The most common low-dose regimen was vincristine and carboplatin. Subjects with TTV score < 20 tend to have normal VA at final review in a significant higher rate, while individuals who scored >30 tend to have severe visual loss of worse than 1.0 LogMAR at last follow-up. However, these patients demonstrated higher rate in vision improvement compared to the other two groups. (Table 3).

Table 2

TTV score	Chemotherapy nl (%)	n2 (%)	Mean Time	Range
<2	9 (30%)	21 (70%)	1.6 yrs	3/12 - 6 yrs
>20 <30	38 (79.16%)	10 (20.83%)	4.2 months	3/7 - 2.1 yrs
>30	36 (100%)	0 (0%)	56.83 days	1/7 - 7/12 yrs

Table 3

TTV score	Normal VA >0.2 N (%)	VA > 0.3 N (%)	Poor VA <1.0 N (%)	Improved N (%)	Stable N (%)	Worse N (%)
<2	21 (70%)	11 (61.11)	1 (11.11)	3 (16.67)	10 (55.55)	5 (27.77)
>20 <30	10 (20.83%)	36 (47.36)	25 (32.89)	15 (19.73)	39 (51.31)	22 (28.94)
>30	0 (0%)		42 (58.33)	19 (26.38)	31 (43.05)	22 (30.55)

Conclusion

Our study demonstrated that the TTV score might be useful in predicting final VA in patients with OPGs. Therefore it could be considered as a substantial factor in the follow-up and management of OPGs and should be taken in consideration by both oncologists and ophthalmologists.

References

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