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Original Article

Ocular characteristics of patients with cerebral palsy

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Abstract

Aim: The purpose of this study is to report the ophthalmic characteristics and treatment results of our cerebral palsy (CP) patients.

Materials and Methods: We explored the charts of 62 CP patients who were followed by our tertiary referral pediatric ophthalmology and strabismus department between 1998-2018 retrospectively. Patient demographics, strabismus type, preoperative deviation, type of surgery, visual acuities (VA), fundus findings, first surgery time, additional surgeries and anomalies and follow-up time were recorded.

Results: Mean age and follow-up time were 34.1 and 42.6 months respectively. Most of the subjects (80.6%) had bilateral VA less than 20/200. The ratio of patients without fixation and follow was 11.2% in right eye and 14.5% in left eye. Fifty eight percent of patients had normal fundus findings. The most leading finding was bilateral optic disc pallor in abnormal fundus patients (33.8%). Fifty-six (90.1%) patients had strabismus (esotropia (ET) (50%), exotropia (XT) (35.5%), vertical deviation (4.8%). Mean horizontal deviation was 25.64±8.41 (8-40) prism diopter (PD) in ET and 25.4±9.39 (12-40) PD in XT. We performed strabismus surgery to 20.9% of patients. The most frequent surgery type was bilateral medial rectus recessions. Mean first surgery time was 35.6 months. Four (30.7%) patients who had previous strabismus surgery needed additional surgery. The most frequent additional surgery types were medial rectus advancement (50%) and inferior oblique myectomy (50%). Prematurity (19.3%), epilepsy (12.9%) and corpus callosum dysplasia and periventricular leukomalacia (12.9%) were the leading encountered additional anomalies.

Conclusions: CP patients are mostly accompanied with esotropia. The management of CP patients may be challenging due to need of additional surgeries.

Keywords: Cerebral palsy; esotropia; exotropia; strabismus; surgery

INTRODUCTION

Cerebral palsy (CP) is a relatively common disorder affecting the motor region of the brain which is non-progressive. CP causes a challenge in permanent posture with lack of independent movement. The exact etiology of CP is unknown but single trauma to the brain induced by antenatal and perinatal factors like prematurity, periventricular leukomalacia (PVL), focal ischemia or hemorrhage or diffuse encephalopathy are the main contributors for this permanent neurological disorder (1).

CP is the leading development abnormality affecting 2 infants of every 1000 newborns around the world whereas this data raises 2 fold to 4.4 infants in Turkey (2,3). Visual disorders usually accompany CP patients addition to orthopedic and neurological impairment. Most common ocular abnormalities in CP are refractive errors (RE), strabismus, nystagmus, amblyopia, observed in %50-90 of CP patients (4-6). Owing to background of evident neurological lesion, the ocular motor control and binocularity are not similar with the patients having ocular disorders without neurologic impairment. In regard to this fact standard ocular management of ocular abnormalities of CP patients is not useful due to unpredictable strabismus surgery results are common (like overcorrections etc.) (7,8). Our aim in this current study was to assess the common ocular and non-ocular disorders of our CP patients and specify our treatment results. We also aimed to compare baseline and final visual acuities (VA) of our subjects.

MATERIALS and METHODS

We collected the data of patients with the diagnosis of CP who were referred to our pediatric ophthalmology and strabismus department of Gulhane Medical School, Ankara-Turkey between January 1998 and January 2018 retrospectively. The study was conducted according to the tenets of the Declaration of Helsinki after confirmation of the institutional ethics committee.

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At first admission, a patient chart was created, and all ophthalmic findings and interventions were recorded. Additional significant extraocular signs or radiological findings were also interviewed with the referral clinic professionals. (pediatric neurology, physical rehabilitation etc.)

We explored and recorded all the available data of patient demographics, strabismus type, preoperative and postoperative deviation, type of surgery, RE, VA (initial and final), fundus findings, first surgery time, additional surgeries, extraocular anomalies and follow-up time. Systemic syndromes or other neurological disorders other than CP were excluded from the study. A detailed ophthalmic examination was achieved including presenting VA, anterior and posterior segment evaluation via standard slit lamp biomicroscope (Zeiss) if applicable. We determined VA by Snellen's chart, Lea Symbol chart or tumbling E chart at 6-meter distance in accordance with age and cooperation of the patient. In uncooperative patients fixation and follow (F/F) characteristics of each eye and also nystagmus and resistance to occlusion patterns were used to guess the VA. Appropriate occlusion therapy was performed in subjects with amblyopia. We measured RE with retinoscopy after 3 times 1% cyclopentolate drops in 5-minute durations achieving a full cycloplegia. We also used autorefractometry (Retinomax; Right Manufacturing, Virginia Beach, VA) in suspected cases for confirmations. Adequate corrective spectacles were prescribed according to age, greatest spherical or cylindrical refractive error of each eye, and also type of ocular misalignment. Anisometropia was defined as presence of >1 diopter difference from fellow eye. We used spherical equivalents (SE), the sum of spherical and ½ of cylindrical error in defining refractive status of patients. All patients were screened for additional anomalies with magnetic resonance imaging and/or computerized tomography to explore retro chiasmal lesions and PVL.

Orthoptic evaluation of the patients was achieved with prism cover test or Krimsky and Hirschberg tests when the first is not possible. Measurement of the angle of deviations were also specified in prism diopters (PD). Any type of ocular muscle restrictions or overaction/underactions were also considered. Head posture and facial symmetry were noted for vertical deviations. Our data was reported as frequency, percentage, mean and standard deviation including min-max. Chi-square test was performed for categorical comparisons via SPSS computer software version 21.0 licensed for Erciyes University.

RESULTS

A total of 62 patients with CP diagnosis were included in this study. Most of our CP cases were male (n:38, 61.2%). At first admission, the mean age was 34.09±18.71 (3-84) months. At final visit, the mean age was 76.76±34.47 (6-168) months. We achieved routine controls and treatment of our patients with a mean follow-up time of 42.61±17.97 (1-84) months. At baseline, most of the subjects (n: 49, 79%) were uncooperative and therefore only F/F were used to guess the VA. Snellen's chart, Lea Symbol chart or tumbling E chart were used to determine the VA in the rest of subjects. Similarly, at final visit 41 subjects (66%) were uncooperative, F/F were used to guess the VA. Snellen's chart, Lea Symbol chart or tumbling E chart were used to determine the VA in 18 patients (29%). The data of VA in 3 (5%) patients were not available in the charts.

Most common initial VA was F+/F+ in 38 patients (bilaterally) of 62 patients. Only 24 of 124 (19.3%) eyes had an initial VA greater than 20/200 while 16 eyes (12.9%) had no light perceptions (NLP) (Table 1).

Table 1. Baseline characteristics of our CP patients

	mean	(min-max)
Age (month)		
Initial visit	34.09	(3-84)
Final visit	76.76	(6-168)
Follow-up (month)	42.61	(1-60)
Angle of deviation (PD)	9.28	(8-40)
Gender	**n**	**(%)**
Male	38	61.3
Female	24	38.7
Visual Acuity		
	Right eye n (%)	**Left eye n (%)**
F+/F+	38 (61.3)	38 (61.3)
F-/F-	7 (11.3)	9 (14.5)
F+/F-	4 (6.4)	2 (3.2)
Counting fingers	1 (1.6)	1 (1.6)
>20/200	12	12
Ocular Findings		
	n	**(%)**
Normal fundus	36	58.1
Esotropia	31	50
Exotropia	22	35.5
Optic disc temporal pallor	15	24.2
Bilateral optic disc pallor/atrophy	6	9.6
Retinopathy of prematurity	4	6.4
Cataract	2	3.2
DVD	2	3.2
4th CNP	1	1.6
Hypertropia+esotropia	1	1.6
IOOA+exotropia	1	1.6
Additional abnormalities		
Prematurity	12	19.4
Epilepsy	8	12.9
Corpus callosum dysplasia	7	11.3
Periventricular leukomalacia	6	9.6
Hydrocephalus/microcephalus	6	9.6
Mental retardation	4	6.4
Head position	2	3.2
Patients with strabismus	56	90.3
Patients with refractive errors	14	22.5

PD; Prism Diopter, F/F; Fixation/Follow, DVD; Dissociated Vertical Deviation, CNP; Cranial Nerve Palsy, IOOA; Inferior Oblique Over Action

The most frequent ocular abnormalities in our CP series were strabismus (n:56, 90.1%), optic disc temporal pallor (n:15, 24.1%), RE (n:14, 22.5%) and nystagmus (n:5, 8%) respectively. In strabismus patients, esotropia (ET) was the leading type of squint (n:31, 55.3%) whereas exotropia (XT) was detected in 22 patients (39.2%). One patient suffered from 4th cranial nerve palsy (CNP). The mean horizontal deviation was 25.64±8.41 (8-40) PD in ET and 25.4±9.39 (12-40) PD in XT. The mean vertical deviation was 10±3.65 (6-14) PD. Other than optic disc (OD) temporal pallor in one eye, 6 (9.6%) patients suffered bilateral OD atrophy/pallor. In patients with RE, the mean of SE was 0.225 in right eye and 0.24 in left eye (Table 1).

Prematurity (n:12, 19.3%), corpus callosum dysplasia (n:7, 11.2%), PVL and hydrocephalus/microcephalus (n:6, 9.6%) were the main detected responsible contributors for the etiology. Apart from prematurity, 4 (6.4%) CP patients suffered retinopathy of prematurity (ROP). ROP cases were treated either laser photocoagulation or only observation where it was appropriate.

Epilepsy was the mostly encountered extraocular neurologic complication affected 8 (12.9%) CP patients (Table 1).

Table 2. Distribution of surgical interventions performed to our CP patients (n: 13)

Initial surgery		
	n	**%**
BMR	8	61.5
BLR	3	23
Lens aspiration	2	15.5
Total	**13**	**100**
Additional surgery (n: 4)		
	n	**%**
IO myectomy	2	50
MR advancement	2	50
Total	**4**	**100**

BMR; Bilateral Medial Rectus Recession, BLR; Bilateral Lateral Rectus Recessions, IO; Inferior Oblique, MR; Medial Rectus

Although a greater number of CP patients had strabismus, only 11 (19.6%) subjects underwent strabismus surgery. The mean age for the first surgery was 36.6±17.49 (10-84) months. The leading surgical procedures for ET and XT were bilateral medial rectus recessions (BMR) (n:8, 72.7%) and bilateral lateral rectus recessions (BLR) (n:3, 27.3%) respectively. We performed bilateral lens aspiration for congenital cataract in 2 patients as an initial intervention (Table 2). In patients with ET, 4 (50%) subjects resulted with under correction while 2 (25%) subjects had overcorrections. Two of 4 patients with no squint after first surgery were ET while the other 2 patients were XT initially. Surgical result of 1 patient with XT was not documented in the chart (Table 3).

A total of 4 patients (30.7%) needed additional interventions in patients who underwent surgery for strabismus. The type of surgeries was medial rectus (MR) advancement for overcorrections of ET in 2 patients and inferior oblique myectomy for inferior oblique over action (IOOA)/4th CNP in 2 patients (Table 2).

Table 3. Post-operative results of the CP patients who underwent strabismus surgery (n: 11)

	n	**%**
Residual esotropia	4	36.4
No squint	4	36.4
Consecutive exotropia	2	18.2
Not documented	1	9.6
Total	**11**	**100**

At final visit, most of our CP patients (n:29, 46.7%) remained the VA of F+/F+ in both eyes. The number of eyes with NLP decreased from 16 to 14. The ratio of patients with an initial VA >20/200 raised from 19.3% to 29% at final visit. The improvement in all VA subgroups were statistically significant (Table 4).

Table 4. Distribution of visual acuities at final visit in our CP patients (n:62) - Initial and final visual acuity comparisons in all eyes (n:124)

Distribution of visual acuities at final visit in our CP patients (n:62)			**Initial and final visual acuity comparisons in all eyes (n:124)**			
	Right eye n (%)	**Left eye n (%)**		**Baseline visual acuity n (%)**	**Final visual acuity n (%)**	**p**
F+/F+	30 (48.3)	29 (46.8)	F+/F+	76 (61.3)	59 (47.6)	0.000
F+/F-	5 (8)	4 (6.4)	F+/F-	6 (4.8)	9 (7.3)	0.000
F-/F-	6 (9.7)	8 (12.9)	F-/F-	16 (12.9)	14 (11.3)	0.000
>20/200	18 (29)	18 (29)	>20/200	24 (19.4)	36 (29)	0.000
Not documented	3 (4.8)	3 (4.8)				

F/F; Fixation/Follow

DISCUSSION

We herein present the clinical data of CP patients with a duration of 20 years. Most of our subjects had low VA at presentation and minority of them gained desirable visual outcome at final visit. Moreover, ocular misalignment, OD pallor and RE were the leading ocular issues hard to cope with to gain a favorable VA both for the clinician and the family. However, the significantly better visual result in our cohort might be due to several reasons. First, as the ages of the subjects getting older, the ability to determine the VA might be improved. Second, the routine ophthalmic visits might have led to better treatment options (refractive error correction, amblyopia treatment, surgical interventions) when needed.

Although overall prevalence of strabismus in normal population is about 3%, ocular misalignment could be seen more frequent ranging 39% from 90% in CP patients (9-12). Consistent with the previous reports, the most common ocular abnormality in our series was strabismus affecting 90.1% of total subjects. Previous reports from our country announced the prevalence of strabismus ranging 50% from 58.7 %and ET was 2-3 folds more frequent than XT in their CP patients (6,11,13,14). Similarly, esodeviation was the leading type of deviation in our series. Adequate explanation with accepted evidence why strabismus is much more frequent in CP patients is lacking. Although it is difficult to locate the exact region of the brain lesion causing strabismus, both oculomotor regions of the brain including cerebellum, especially significant harm to the white matter, seems to be possible contributors (15).

Furthermore, owing to CP has variable etiology ranging hypoxic–ischemic encephalopathy to periventricular white matter lesions resulting neurodevelopmental damage which could affect both ocular motor and visual pathways may cause visual impairment (VI) independent of ocular abnormalities (11). Addition to this, PVL was linked to VI and unfavorable functional outcome owing to adverse impact on visual associated areas in the brain (11,16,17). Despite appropriate treatment was performed, this aforementioned fact could be the reason for our high percentage of CP cases remained with VI. In a recent report, the authors argued that transsynaptic degeneration of the neurons in central visual pathways could be responsible for the typical retina and OD presentation (pallor, atrophy etc.) seen in CP patients indicating that most patients in this cohort had some degree of optic nerve atrophy (11, 18, 19). A greater number of patients (n:21, 33.8%) had this distinctive optic disc appearance supporting this argument described above.

Surgical management of ocular misalignment in CP patients is difficult. First, appropriate measurement of the deviation is challenging due to most of the subjects having poor cooperation to the examination before surgery. Second, standard nomograms for surgery planning is not much effective compared to normal population. Unpredictable surgical results (overcorrection/under correction) are not uncommon (7,8). Even though modification of lessening 1mm for each muscle from standard tables for surgery planning in ET, overcorrection rates were found to be significantly higher at final visit with MR recessions (8). Although we had only 8 patients who underwent BMR, over correction rate was 25% after first surgery which was consistent with the study of Ma and co-workers' final result (8). Unfortunately, we did not have the data of final result of overcorrected patients who underwent additional MR advancement procedures to compare. An interesting result of our study was that most of patients (50%) with ET were under corrected after first surgery in contrary to published reports.

Other than surgery, botulinum toxin injections to bilateral MR were reported to be safer and having more predictable results than surgery with a high success rates (61.4%) for ET in CP (20). In a report of Ma et al comparing the surgical results of BLR in CP patients with XT, the authors found indifferent surgical responses for this procedure with non-CP subjects (21). Two of 3 CP patients with XT in our study achieved surgical success comparable with the previous report despite the limited number (only 3) of patients (21).

Most of the studies focus on horizontal deviations in CP while in Medline search, we could not find any published data about management of vertical deviations (IOOA/4th CNP) in CP. We had 2 CP subjects with vertical deviations who were well-treated with surgery. We think similar responses to surgery in CP patients with vertical deviations can be achieved without the need of any modifications in surgery nomograms like XT.

This study has limitations. First, one-center, retrospective and non-comparative design of the study limits the generalization of the results to all CP patients. Second, objective measurement of VA was not possible in all patients owing to higher rate of subjects with low VA. Third, complete assessment of sensory tests including stereopsis, amblyopia and binocular function screening could not be achieved due to low cooperation. Moreover, we are aware that the number of patients who underwent strabismus surgery was not enough to make perfect comparisons with previous reports. Additionally, we could not specify the clinical types of CP (spastic, dyskinetic, ataxic) which could have changed our results. However, we think presentation of CP patients with unfavorable VA both initially and finally will be placed in a valuable position in literature. Our high rate of under corrections to BMR surgery in ET and well-treated vertical deviations were distinctive results of this current study.

CONCLUSION

In conclusion, we report a large number of CP cases with a long period of time (20 years). Most of the cases in our series were with strabismus (>90%) and having OD abnormalities (33.8%). Despite the fact that most of our CP individuals achieved normal ocular alignment after strabismus surgery, unfortunately the functional outcome of patients was not satisfactory at final visit.

Conflict of interest : The authors declare that they have no competing interest.

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