The Bell's phenomenon in newborns

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ABSTRACT

Objective: This study was undertaken to detect the course of Bell's phenomenon development in full and pre-term babies.

Methods: Three hundred healthy newborns (150 full term and 150 pre-term) born at Prince Rashid Bin Al-Hasan military hospital from the first of January 2000 to 31st of December 2000 in Irbid in the north of Jordan, were randomly selected and included in the study. Bell's phenomenon response was graded according to the movement of the eye as: 0 = no movement, 1 = minimal movement (the pupil still visible), 2 = full movement (the pupil is covered by the eyelid). The Bell's Phenomenon Scoring System (BPSS) was chosen to score the eye movement. The newborn was examined during the first week of life and then repeated at 4 weekly intervals, for 5 months for the full term babies and for 2 months for the pre-term babies.

Results: In the full term group only 25/150 (16.7%) of the newborns showed full response (BPSS-2) during the first week of life and this number increased to 133/150 (88.7%) at the age of 5 months. None of the pre-term group showed the Bell's phenomenon response (BPSS-0) during the first week of life and only 21/150 (14%) had full response (BPSS-2) at 4 weeks. At the age of 2 months approximately 51/150 (34%) developed full response (BPSS-2).

Conclusion: The development of Bell's phenomenon reflex reflects the maturation process of the brainstem and the extraocular muscles related to eye elevation. It should be considered as part of the routine tests undertaken by every pediatrician and ophthalmologist during the neuro-ophthalmic examination in newborns.

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Bell's phenomenon (BP) is the upward deviation of the eye during attempted eyelid closure. It is an important physical sign in clinical medicine, which may help to explain or localize the cause of ocular disease and lead to the diagnosis of systemic disease. It was first described by Sir Charles Bell in 1823 in the presence of a unilateral lower motor neuron lesion of the seventh cranial nerve.[1] It is an ocular polysynaptic somatic reflex that includes eyelids and extraocular muscles,[2] such that the eyes typically roll upward and outward when efforts are made to close the eyelids against resistance. The neural mechanism for this integrated movement is unknown yet, but involves brainstem pathways between the seventh nerve nucleus in the pons and the third nerve complex in the rostral midbrain.[1,3]

Snir et al[4] evaluated the development and course of BP in premature and normal neonates. They found that no preterm infants exhibited BP in the first week of life and only 29.6% exhibited a weak to full reflex at 8 weeks of age, while in the full term infants 35.7% demonstrated a mature reflex in the first week and 97.3% at age 16 weeks. Hall[5] and Hoyt et al[6] in their studies in normal neonates noted that transient disturbances of the vertical gaze are common.

In our study, we tried to detect the development of BP in 300 newborns (150 full term and 150 preterm babies) in our region in the north of Jordan.

Methods. One hundred and fifty premature babies (71 (47.3%) males and 79 (52.7%) females)

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who were born between 28 and 36 weeks of gestation were randomly selected and involved in the study. Another group of 150 mature babies (89 (59.3%) males and 61 (40.7%) females) who were born between 37 and 42 weeks of gestation were also randomly selected.

They were born at Prince Rashid Bin Al-Hasan military hospital from the first of January 2000 to 31st of December 2000 in Irbid in the north of Jordan. BP was assessed by opening the baby's eyelids widely while the baby was attempting to close them forcibly. This was repeated by holding both eyelids open against resistance. The BP score system (BPSS) was chosen to score the eye movements, and it was graded from 0-2 as follows: 0: no movement, 1: slight movement (pupil visible), 2: full movement (pupil invisible, namely, covered by the eyelid). The ocular examination was carried out on the first week of life for all babies and repeated at 4 weekly intervals for babies who showed weak response, or none at all, until they showed the full response. On each visit, each baby had body weight, head circumference, length measurements and neurological reflexes including pupillary light, oculovestibular and Doll's eye movements recorded on his growth chart. The babies were followed up until they developed full BP. Any child who had a problem that may affect the development of BP was excluded from the study. This includes maternal diseases such as diabetes mellitus, myasthenia gravis, myotonic dystrophy, renal transplant, malignancies, hypertension, hyperparathyroidism, Grave's disease, endemic goiter, drug abuse and cyanotic heart diseases, and baby-related problems like cesarian section, hydrocephalus, convulsions, chromosomal abnormalities, intraventricular hemorrhage, labor asphyxia, neurological disorders and Apgar score less than 7. Eye related congenital anomalies like eyelid coloboma, extra-ocular muscles disease, for example, Duane retraction syndrome or orbital anomaly were excluded. And finally any baby who was lost to follow was excluded also.

Results. In the full term group, 150 babies (89 (59.3%) males and 61 (40.7%) females) were included in the study. The gestational age ranged from 37-42 weeks with a mean of 38.7 weeks, mean birth weight of 3264.6 grams and head circumference of 35.4 cm. **Table 1** demonstrates the BP scores on each examination in the full term group. While 133 (88.7%) of 150 full term babies had full BP at the age of 5 months, 8 (5.3%) babies had a weak response, and 4 (2.7%) babies had no response.

In the second group 150 premature babies (71 (47.3%) males, 79 (52.7%) females) were included. The gestational age ranged between 28-36 weeks of gestation, and their birth weight between 750-2500 grams. **Table 2** demonstrates the BP scores on each examination in the premature group. In summary, at the age of 2 months 48 (32%) premature babies showed BPSS-2, 15 (10%) BPSS-1, 3 (2%) variable responses and 84 (56%) no response at all.

Discussion. The Bell's phenomenon mechanism involves the seventh cranial nerve nucleus in the pons and the third cranial nerve nuclear complex in the rostral midbrain and the interconnecting pathways; however, the exact mechanism was unknown.[1] In a study carried out by Cogan[7] on normal volunteers, 132/156 eyes deviated upwards or upwards and laterally, and the other eyes showed variable responses. In 1984, Francis and Loughhead[8] reported their findings in 508 normal patients who presented consecutively and were examined for their BP and for lagophthalmos. They

Table 1 - Bell's phenomenon score (BPSS) on each examination in the full term babies.

Examination (age)	BPSS-0 n (%)	BPSS-1 n (%)	BPSS-2 n (%)	Variations n (%)	TOTAL
1st (One week)	84 (56%)	36 (24%)	25 (16.7%)	5 (3.3%)*	150
2nd (4 weeks)	51 (42.5%)	45 (37.5%)	24 (20%)	-	120
3rd (8 weeks)	16 (16.7%)	28 (29.2%)	52 (52.4%)	-	96
4th (12 weeks)	8 (18.2%)	17 (38.6%)	19 (43.2%)	-	44
5th (16 weeks)	6 (24%)	11 (44%)	8 (32%)	-	25
6th (20 weeks)	4 (23.5%)	8 (47.1%)	5 (29.4%)	-	17
TOTAL	**4 (2.7%)**	**8 (5.3%)**	**133 (88.7%)**	**5 (3.3%)**	**150**

*Abnormal response in the form of either inward or inward and downward movement of the eye in the first exam.

Table 2 - Bell's phenomenon score (BPSS) on each examination in the premature babies.

Examination (age)	BPSS-0 n (%)	BPSS-1 n (%)	BPSS-2 n (%)	Variations n (%)	TOTAL
1st (One week)	150 (100%)	0 (0%)	0 (0%)	0 (0)	150
2nd (4 weeks)	108 (72%)	19 (12.7%)	21 (14%)	2 (1.2)	150
3rd (8 weeks)	84 (66.1%)	15 (11.8%)	27 (23.1%)	1 (0.8)	129
TOTAL	**84 (56%)**	**15 (10%)**	**48 (32%)**	**3 (2%)**	**150**

demonstrated a good deal of variability not only in the amount but also in the type of BP response in normal patients. They found variability in the type of BP in 21%; none had a negative response, 7% had a minimal upward response, and 8% of patients showed downward response. Ferrer[9] in 1973 studied the incidence of BP among 379 normal children from birth to age 5 years. From 124 babies, only 13 (10.5%) had normal upward gaze response up to the age of 2 months, 23.6% at the age of 4 months and 33.3% at the age of 8 months. A group of 180 children over the age of 5 years showed a normal BP in 88.9%. In 1993 Snir et al[4] evaluated the development and the course of BP in premature and normal neonates. They found that in the full term babies, 35.7% revealed a positive BP on the third day of life, 50% at the age 4-8 weeks.

In our study, 20% of the full term babies showed a positive BP on the first week of life, 36% at the age of one month, 70.7% at 2 months and 83.3% in the 4th month of life. The discrepancy between the results of our study, Snir's et al[4] and Ferrer's[9] may be attributable to the difference in the size of the studied samples. At the age of 5 months, 92% of full term babies had a positive BP, which was slightly more than that of Ferrer's group that was examined over the age of 5 years. This may be due to number of examined babies, which were more in Ferrer's[9] group. Snir et al[4] also examined another group of 27 premature babies, and they found a negative BP in the first week of life and at the age of 2 months only 29.6% showed weak to full BP response. In our findings of the premature group (150 babies), none had BP response in the first week of life, supporting the findings reported by Snir et al.[4] However, at the age of 2 months, 44% of premature babies showed BP response, which is more than that of Snir's et al at the same age.

In our study, there were variations in BP response as either inward or inward and downward deviation of the eyes in 3.3% in the full term and 2% in the premature babies. These percentages were less than that found by Francis and Loughhead[8] and Snir et al,[4] but their findings might be transient disturbances of the vertical gaze which is common in normal neonates as Hall[5] and Hoyt et al[6] noted in their studies.

Our findings of gradual increase in the appearance of BP in full term and premature babies supports the previous assumption that BP has a longitudinal neurological development and is part of the normal neurological maturation process. They also support the assumption that maturity of the brainstem is a prerequisite for the normal appearance and the variability of BP. It should be considered as part of the routine tests undertaken by every pediatrician and ophthalmologist during the neuro-ophthalmic examination in newborns.

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