

Bell's Phenomenon during Screening Examination for Retinopathy of Prematurity

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Purpose: Bell's phenomenon (BP), which may disturb screening examinations for retinopathy of prematurity (ROP), is known to present infrequently in premature babies. Stress associated with the examinations can influence expression of BP. The authors of the present study evaluated BP during examinations for ROP.

Methods: The present study included 102 eyes of 51 premature babies. Expression of BP was assessed at 3 steps of the examination in the following order: after insertion of a speculum, after illumination of an indirect ophthalmoscope and after scleral depression. The relationship between the expression of BP and the gestational age at the examination was analyzed in each step of the examination.

Results: The frequency of BP after the speculum insertion and the illumination was 77% to 92% in infants 32 weeks of age or younger, and decreased significantly to 16% to 57% in infants 42 weeks of age or older ($p < 0.005$). BP after the scleral depression had no significant association with the gestational age. Frequency of BP increased significantly as the steps of the examination proceeded ($p < 0.01$).

Conclusions: BP was frequent in premature infants during ROP examination in spite of neurological immaturity. The examiner should take BP into consideration, which frequently occurs in younger infants.

Key Words: Bell's phenomenon, Retinopathy of prematurity

Bell's phenomenon (BP) was reported by Bell in 1823 [1] as an upward deviation of the eye during attempted eyelid closure in the presence of a lower motor neuron defect of the facial nerve. Although the exact mechanism is unknown, Hiraoka [2] suggested the mesencephalic reticular nucleus may play an important role in integrating 2 patterns of movement. BP was observed in more than 80% of normal adults [3,4]. The frequency of BP was, however, reported as low as 10.5% in newborn infants and increased gradually during the first year of life [3,5]. Ferrer [3] reported that 23.6% of infants showed a normal BP at the age of 4 months and 52.3% at the age of 1 year. Snir et al. [5], on the other hand, reported 35.7% of full-term babies revealed a positive BP on the third day of life and 97.3%

in the fourth month of life. In premature babies, no infants exhibited BP during the first week of life and only 8 out of 27 (29.6%) showed BP at the age of 8 weeks.

The previous reports assessed BP by opening the eyelids with the examiner's fingers [3,4]. As BP is a variable phenomenon elicited by a voluntary movement or attempt to close the eyelids, the frequency of BP could be influenced by the methods that were used to assess the movements of the eyeballs. Consequently, BP is expected to be induced in an examination for retinopathy of prematurity (ROP), which can be stressful enough to make a baby attempt to close the eyelids forcefully.

Manifestation of BP was reportedly related with conjunctival injury during ROP examination [6]. The conjunctival injury or swelling may disturb the following examination and laser treatment. The purpose of the present study was to evaluate the relationship between the expression of BP and the examination for ROP.

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Materials and Methods

Fifty-one premature infants without neurologic defects or diseases were enrolled in the present study. Birth weights, gestational age at birth, and medical as well as neurological status were reviewed for each baby. The examinations for ROP conformed to the guideline by the American Academy of Pediatrics (revised in 2001). During the examinations, the amount and direction of BP were evaluated. The protocol was approved by the institutional review board of the Pusan National University Hospital and the present study was compliant with the principles of the Declaration of Helsinki.

A serial number was given to each baby. At the first examination, the babies with odd numbers had the right eye (the preceding eye) examined before the left eye (the following eye), and for the babies with even numbers, the left eye was the preceding eye and examined first. At the next examination, the following eye of the previous examination was examined first before the other eye. Topical anesthetic was not given.

The BP was evaluated at 3 steps of each examination: 1) after the insertion of the speculum, 2) after the examination of the posterior pole with the illumination of the light from the indirect ophthalmoscope, and 3) after the scleral depression. The last step was immediately after identifying the ora serrata at 3 and 9 o'clock. If necessary, further examination for other clock hours was performed. The amount of BP was graded by 2 observers as follows: 0, no response; 1, minimal response, or the center of the cornea is seen, and 2, full response or the center of the cornea is not seen (Fig. 1). All examinations were performed by the same ophthalmologist. The BP was evaluated for more than 3 seconds. If eye movements did not continue for 3 seconds and have a steady direction, the movements were excluded to rule out wandering eye movements. The speculums and depressors of the same size and the same indirect ophthalmoscope were used.

The gestational ages at the examination were divided

into 5 periods: 1) 32 weeks or younger, 2) 33 to 35 weeks, 3) 36 to 38 weeks, 4) 39 to 41 weeks, and 5) 42 weeks or older. If a baby was examined more than once in 1 period, the first examination was used for the analysis.

The baseline characteristics were analyzed. The relationship between BP and the gestational age at the examination was evaluated by χ^2 -test with SPSS ver. 12.0 (SPSS Inc., Chicago, IL, USA). To avoid a bias by multiple counts, BP at 36 to 38 weeks of gestational age was selected and analyzed for the association with the steps of the examination, the precedence, or the laterality of the eyes by χ^2 -test.

Results

A total of 102 eyes from 51 premature infants were enrolled in the present study. The mean gestational age at birth was 31.0 ± 2.8 weeks (range, 25 to 37 weeks) and the mean birth weight was $1,465.8 \pm 450.2$ g (range, 643 to 2,830 g). The birth weight was correlated with the gestational age. Supplemental oxygen was given to 28 babies; 16 had a respiratory distress syndrome. Cardiovascular abnormalities were diagnosed in 7 babies. Other risk factors for ROP included sepsis in 3 and transfusion in 2 babies. No infant had intraventricular hemorrhage, periventricular leukomalacia or other cerebral problems that may cause abnormal eyeball movements. During the follow-up, significant ROP developed in 3 eyes of 2 babies. All regressed after laser treatment.

ROP examinations were performed 153 times in total. The average number of the examinations for each infant was 3.0. The number of ROP examinations analyzed in each period was 13 in 32 weeks of age or younger, 38 in 33 to 35 weeks of age, 50 in 36 to 38 weeks of age, 27 in 39 to 41 weeks of age and 19 in 42 weeks of age or older.

BP was usually transient and the location of the eyeball returned straight from up-deviation in several seconds. The frequency of BP after the insertion of the speculum decreased significantly as the gestational age at the examination increased from 85% (8% in grade 1, 77% in grade

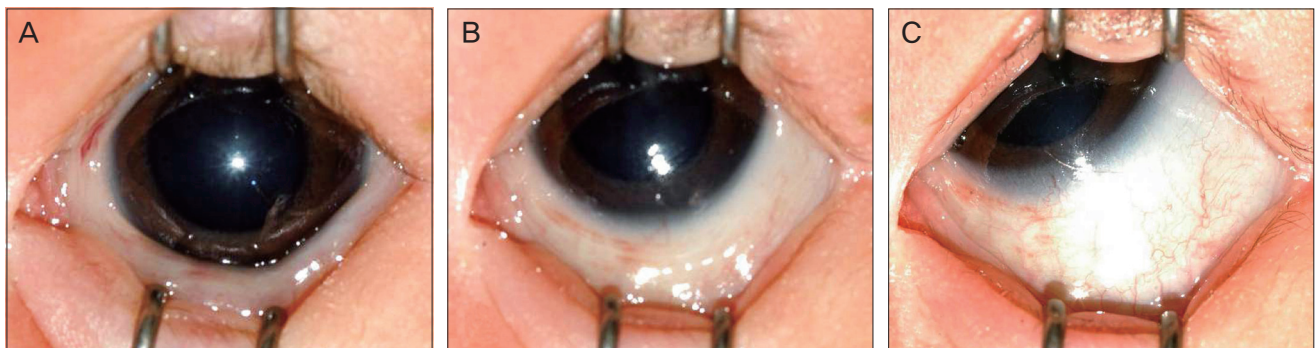


Fig. 1. Grading of Bell's phenomenon: (A) grade 0, no response; (B) grade 1, minimal response, or the center of cornea is seen; (C) grade 2, full response or the center of the cornea is not seen.

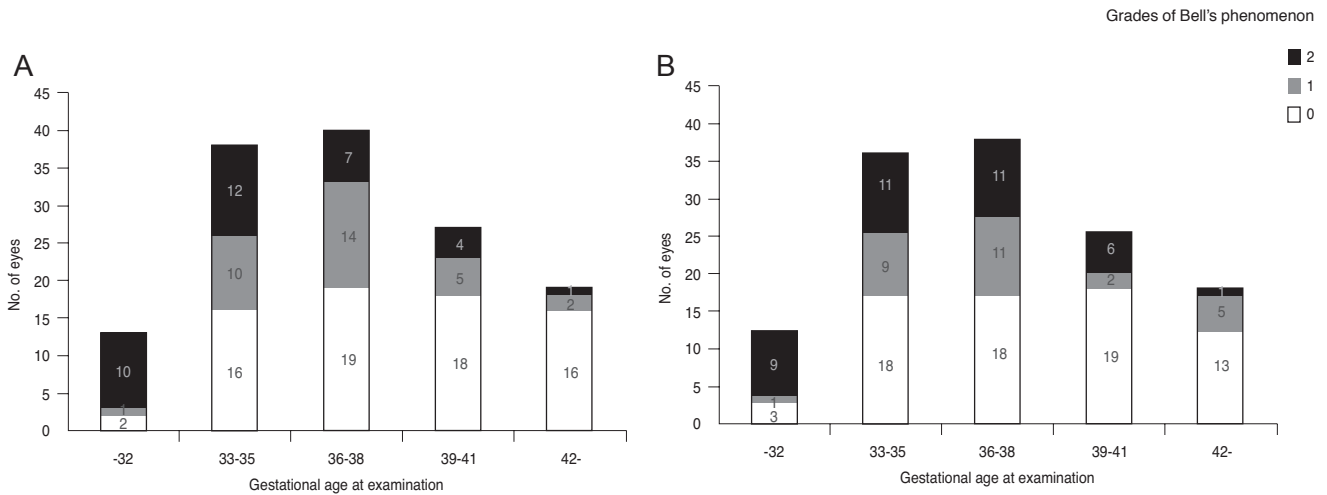


Fig. 2. Frequency and grade of Bell's phenomenon after insertion of speculum decrease significantly according to gestational age at examination for retinopathy of prematurity of (A) right eyes ($p < 0.001$) and (B) left eyes ($p = 0.005$).

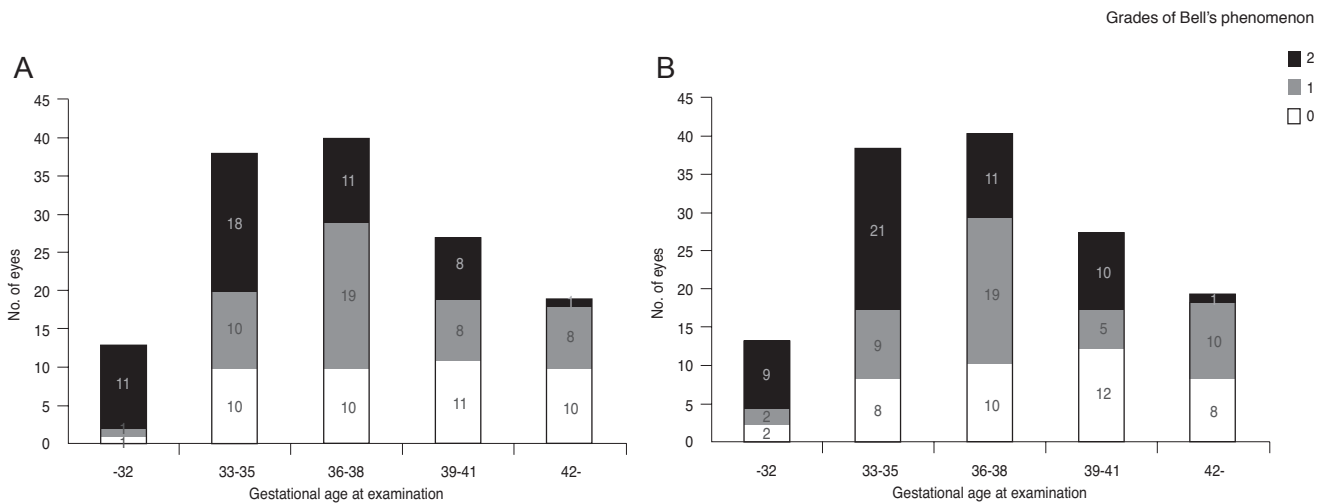


Fig. 3. Frequency and grade of Bell's phenomenon after illumination of indirect ophthalmoscope decrease significantly according to gestational age at examination for retinopathy of prematurity of (A) right eyes ($p < 0.001$) and (B) left eyes ($p = 0.001$).

2) in the right eyes and 77% (8% in grade 1, 69% in grade 2) in the left eyes at 32 weeks of age or less to 16% (11%, 5%) and 32% (27%, 5%) at 42 weeks of age or more ($p < 0.001$ and $p = 0.005$, respectively) (Fig. 2). Similarly, the expression of BP after the illumination of the indirect ophthalmoscope decreased significantly from 92% (8%, 84%) and 85% (16%, 69%) to 47% (42%, 5%) and 58% (53%, 5%) ($p < 0.001$ in the right eyes and $p = 0.001$ in the left eyes, respectively) (Fig. 3). No significant association was found between the gestational age at the examination and the expression of BP after the scleral depression (Fig. 4).

The expression of BP at 36 to 38 weeks of age increased significantly as the steps of the examination progressed ($p < 0.001$). No significant difference regarding the laterality or the precedence of the eye was found (Figs. 5 and 6).

Discussion

BP is the deviation of an eye, which accompanies an attempted closure of the eyelids. Ferrer [3] reported 78% of normal subjects over 5 years of age had BP. Francis and Loughhaed [4] observed BP, including a small up-response in all 508 normal patients and reported much variability in the amount and type of BP response.

BP was thought to be uncommon in premature infants because of a neurological immaturity [5]. Snir et al. [5] reported the incidence of BP in premature babies increased gradually after birth and only 8 out of 27 (29.6%) showed BP at the age of 8 weeks. The results appear contradictory to the results in the present study which showed BP was present after scleral depression in more than 50% of the

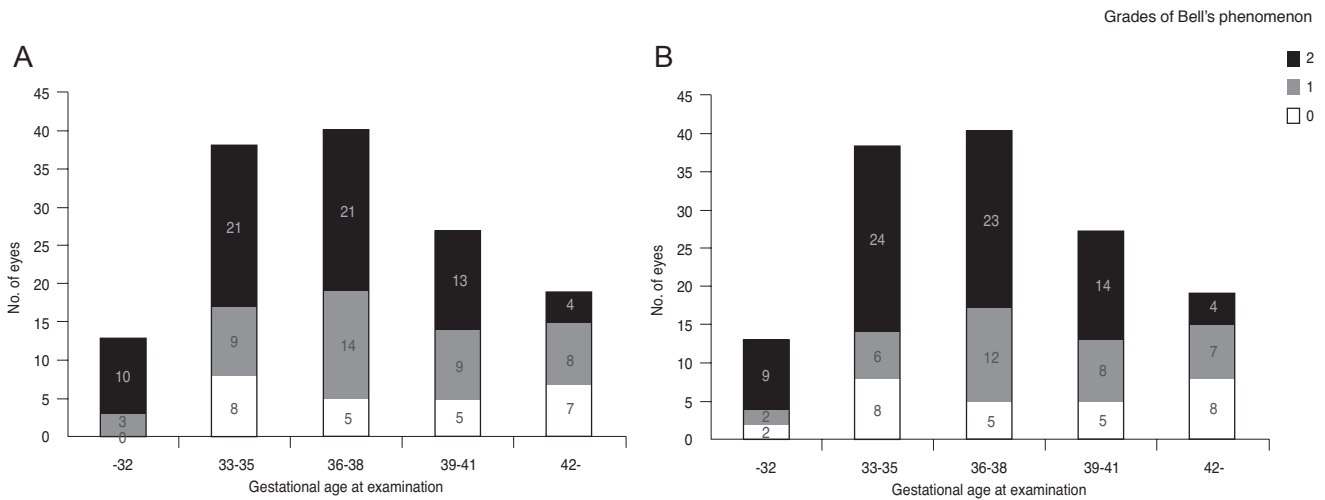


Fig. 4. Frequency and grade of Bell's phenomenon after scleral depression do not show a difference according to gestational age at examination for retinopathy of prematurity of (A) right eyes ($p = 0.080$) and (B) left eyes ($p = 0.075$).

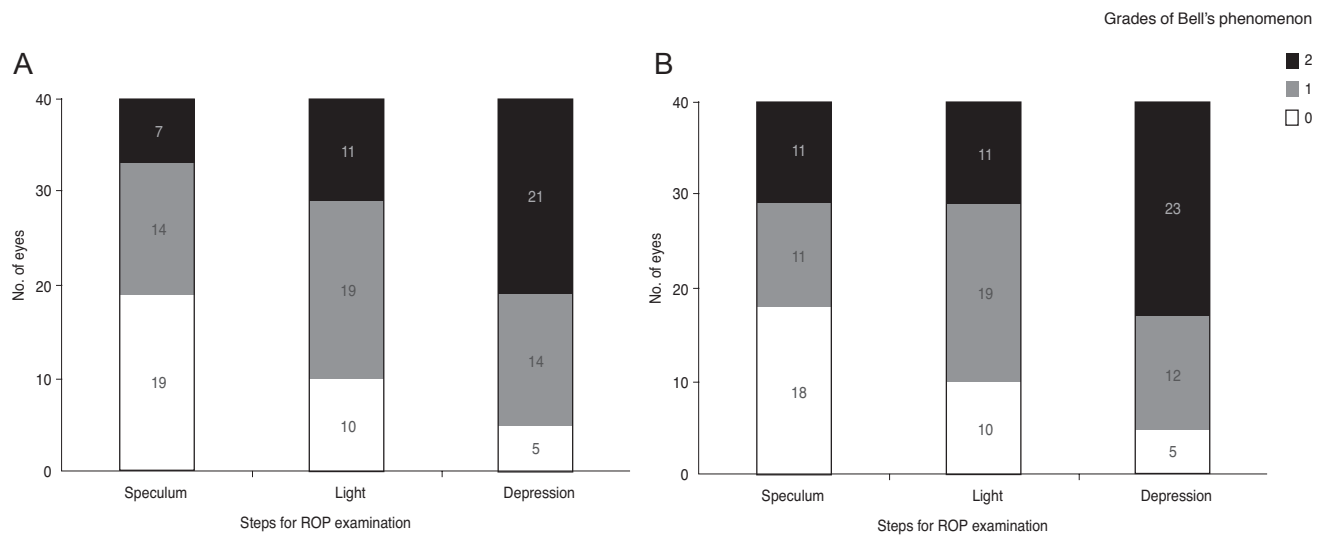


Fig. 5. Frequency and grade of Bell's phenomenon increase significantly according to progression of the examination in premature infants with a gestational age of 36 to 38 weeks at examination for retinopathy of prematurity (ROP) of (A) right eyes ($p < 0.001$) and (B) left eyes ($p < 0.001$). No difference was found between the right and the left eyes.

premature babies regardless of their age. In addition, more than 70% of the premature babies at the gestational age of 32 weeks or less showed BP only after the insertion of the speculum. Furthermore, the frequency of BP decreased as the babies grew. The movements of the eyeballs were assessed because BP was thought to be possibly associated with abnormal alignments which have a high incidence in neonates [7]; however the misalignments are usually horizontal [7,8].

The differences in the condition under which BP was assessed would explain the contradiction. Voluntariness appears important for the expression of BP. Paez et al. [9] found the elevation of the eyeballs associated with BP was not shown during general anesthesia with or without mus-

cle relaxants. The voluntariness can be achieved simply by a request to adults, whereas obtaining the cooperation from newborn infants for the assessment of BP is difficult. Snir et al. [5] examined BP by opening the eyelids gently with the fingers and defined the attempt to close the eyelids as the resistance against holding them open. In the present study, the voluntariness could not be assessed as the speculum was inserted. The insertion of the speculum was thought more stressful than the gentle opening by the fingers, and could make the baby attempt to close the eyelids forcefully. The higher rate of BP could result from such voluntary attempts in the present study. Therefore, the low expression of BP in premature infants of the previous studies is possibly due to the weak voluntariness to close

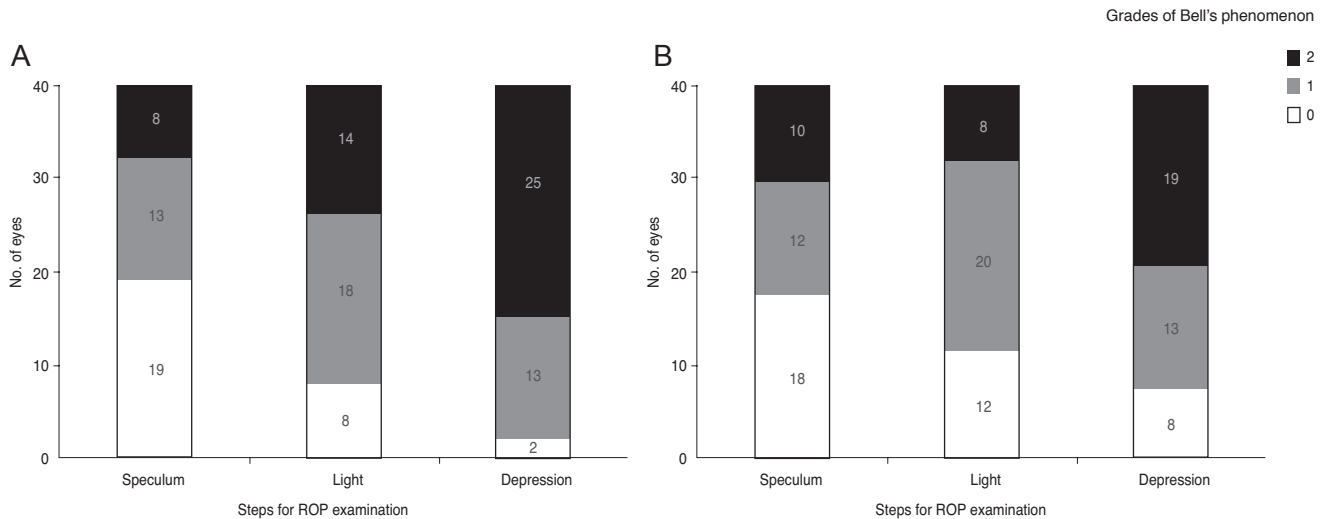


Fig. 6. Frequency and grade of Bell's phenomenon increase significantly according to progression of the examination in premature infants with a gestational age of 36 to 38 weeks at examination for retinopathy of prematurity (ROP) of (A) preceding eyes ($p < 0.001$) and (B) following eyes ($p < 0.001$). No difference was found between the preceding and the following eyes.

the eyelids rather than neurological immaturation.

Evidence for the authors' hypothesis would be the expression of BP increasing significantly with the progression of the examination. The latter steps for ROP examination included the previous steps fundamentally. Therefore, the stress of the third step was actually the summation of the stresses from each step; the speculum, the light of the indirect ophthalmoscope and the scleral depression. Consequently, BP could be induced at a higher rate in the latter steps. The stress did not persist to affect the examination of the other eye, because BP was usually transient, and no differences were found regarding the laterality or the precedence of the eyes.

The results from the present study implicate the ROP examination was more stressful in the more premature infants. The size of the speculum could be a factor causing a significant stress, because speculums of the same size were used, which were relatively large for a younger infant. In addition, the relatively narrow eye fissures of the Asians could be related with the higher rate of BP. The scleral depression is thought as a strong enough stress to induce BP in premature infants regardless of the age.

The examiner may experience difficulty when examining the inferior retina of the eye showing strong BP, because of the usual upward rotation. Park et al. [6] reported the manifestation of BP was related with conjunctival injury during ROP examination. They explained the conjunctival injury was caused by forceful depression to manipulate the eyeball inferiorly. Even if conjunctival injury is not caused, the examination can be prolonged and result in the edema of the conjunctiva. The following examination and laser treatment will be disturbed by the conjunctival injury or swelling, from which misdiagnosis or undertreatment

may result.

In summary, the incidence of BP in premature infants during ROP examination was higher in the present study than in the previous reports. Because BP manifests more frequently in younger infants, the examiner should be especially attentive on examination.

Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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