PURPOSE: Pierre Robin sequence (PRS) is characterized by micrognathia, glossoptosis, and airway obstruction. Respiratory status and jaw development are often focused as outcomes in the treatment of PRS. Growth and development can be regarded as true outcomes in the treatment of congenital anomaly. In the present study, we focused on body weight and development of the patients with PRS treated at National Center for Child Health and Development, Tokyo, Japan to elucidate these outcomes.

METHODS: Our present strategy for PRS patients include conservative treatments such as positioning, nasal airway, and noninvasive positive pressure ventilation. Surgical treatments were provided for patients who are intractable to these measures and included tongue-to-lip adhesion and tracheostomy. Patients who were diagnosed as PRS were collected from our database. Patients were included if they required treatment for airway obstruction during infancy and data on body weight and development later than 2 years old were available. Retrospective chart review was performed. Body weight was evaluated based on Japanese norms, and developmental delay was deemed present if IQ/DQ was lower than 70 or enrolling in special needs classes.

RESULTS: Seventeen patients were included. Median length of follow-up was 11 years (range: 2–19 years). Eight patients were isolated PRS, four patients were syndromic (two patients with Stickler syndrome, one patient with Cornelia de Lange syndrome, and another with chromosomal abnormality), and five patients were associated with other congenital anomalies. Surgical intervention for airway obstruction was required in four patients (two requiring tongue-to-lip adhesion, and three requiring tracheostomy, with one patient undergoing both). The number of patients with body weight heavier or equal to Japanese average was 10 of 17 patients at birth, declined to 1 of 13 during neonatal period (1–2 months old), and 5 of 14 during toddler to pre-school ages (2- to 5-year-olds), and 6 of 8 during school ages and thereafter. There was no significant difference in the transition of body weight between isolated and non-isolated PRS. Developmental delay was present in six patients.

DISCUSSIONS: Rate of body weight increase declined dramatically during neonate. Catch-up growth was observed during infancy, but body weight in more than half of the patients was lighter than Japanese norms during their toddler to pre-school age. Recently, better increase in body weight has been reported with mandibular distraction osteogenesis and baton plate. The results of the present study pose a need for a more aggressive approach than our present protocol to enable better growth and development for the patients with PRS.

REFERENCE:
1. Paes EC, de Vries IAC, Penris WM, et al. Growth and prevalence of feeding difficulties in children with Robin sequence: a retrospective cohort study. Clin Oral Investig. 2017;21(6):2063–2076.
mild ($1 < \text{MMEG} < 2$), moderate ($2 < \text{MMEG} < 3$), and severe ($\text{MMEG} = 3$). Demographic variables were analyzed by chi-square test, one-way ANOVA, and student $t$-test.

RESULTS: Of the 466 patients who underwent an eyelid procedure, there were 103 patients who had postoperative edema that was evaluated to have an MMEG of 1 or higher. An estimated 35 patients had mild edema ($1 < \text{MMEG} < 2$), 44 patients had moderate edema ($2 < \text{MMEG} < 3$), and 24 patients had severe edema ($\text{MMEG} = 3$). The results of the chi-square analysis demonstrated that there was a significant association between ethnicity and degree of postoperative swelling (**$P < 0.0001$). The one-way ANOVA indicated that there was a significant difference in maximum mean edema grade (MMEG) across the racial and ethnic populations included in the study (*$P < 0.05$). In addition, the data indicated that Asian patients ($1.98 \pm 0.81$) had significantly higher edema grades than White patients ($1.35 \pm 0.78$) and when compared using student $t$-test (***$P < 0.001$).

CONCLUSIONS: In patients undergoing an eyelid procedure, the degree of postoperative eyelid swelling was significantly associated with ethnicity. In addition, patients of the Asian population demonstrated significantly higher grades of eyelid edema compared with those of the White populations.