# SURGICAL RISK FACTORS IN LARSEN'S SYNDROME

K. KARAKAS[1], E. F. PERCIN[2], S. PERCIN[1]

**The authors report on a child with typical Larsen's syndrome with some rare findings such as mixed-type hearing loss and with some potentially fatal operative risks including laryngomalacia and cervical instability. A few deaths with Larsen's syndrome have been reported associated with various fatal risks such as spinal instability. Therefore, laryngomalacia and several other potentially fatal risks are presented in this report as awareness may prove essential to orthopedic surgeons.**

**Keywords** : Larsen's syndrome ; multiple joint dislocations ; laryngomalacia ; cervical instability.
**Mot-clés** : syndrome de Larsen ; luxations multiples ; laryngomalacie ; instabilité cervicale.

## INTRODUCTION

Larsen's syndrome is a structural genetic disorder, autosomal dominant, autosomal recessive or sporadic (11). McFarland first described an individual case in 1929 (4, 11). In 1950, Larsen *et al.* described the components of the peculiar facies with a flat face (hypertelorism, prominent forehead and depressed nasal bridge), multiple congenital large joint dislocations and foot abnormalities (pes equinovarus or valgus) as a distinct syndrome in six cases (7, 8). Other features of Larsen's syndrome are hand abnormalities (accessory carpal bones, short terminal phalanges, long cylindrical fingers, shortened metacarpals, triangular terminal phalanges of the thumb) (7).

In the case reported here, we witnessed severe intraoperative respiratory problems related to isolated laryngomalacia. It has been previously reported that some anomalies such as spinal instability (1, 2, 12), mobile arytenoid cartilage (7), and tracheomalacia (3, 10, 16), laryngotracheomalacia (13) and laryngotracheobronchomalacia (17) in children with Larsen's syndrome have caused serious problems and even death during operation. We therefore thought it worthwhile to report this case.

## CASE REPORT

S. I., a 32-month-old girl, was born at term. The mother and father were first-degree relatives, 24 and 30 years old respectively. During the pregnancy her mother was not exposed to agents known to be mutagenic or teratogenic, and she did not recall bleeding or any serious infectious diseases. The family history revealed that there had been two miscarriages and one child with anencephaly who died at 32 weeks gestation. The girl had normal intelligence and could speak but not walk. The case history showed that laryngeal stridor and various deformities had been present since birth.

The patient was 10 kg in weight and 67 cm in length. Head and chest circumferences were 47 cm and 52 cm, respectively. On inspection, the typical flat face was remarkable ; the girl also had a depressed nasal bridge, wide-spaced eyes, prominent forehead, micrognathia and a short neck. Inspiratory stridor was heard. There were rales and

[1] Cumhuriyet University, Faculty of Medicine, Department of Orthopedic Surgery and Traumatology, Sivas, Turkey.
[2] Cumhuriyet University, Faculty of Medicine, Department of Medical Biology and Genetics, Sivas, Turkey.
Correspondence and reprints : K. Karakas, Cumhuriyet Üniversitesi Tıp Fakültesi, Ortopedi ve Travmatoloji Anabilim dalı, 58140 Sivas, Turkey.

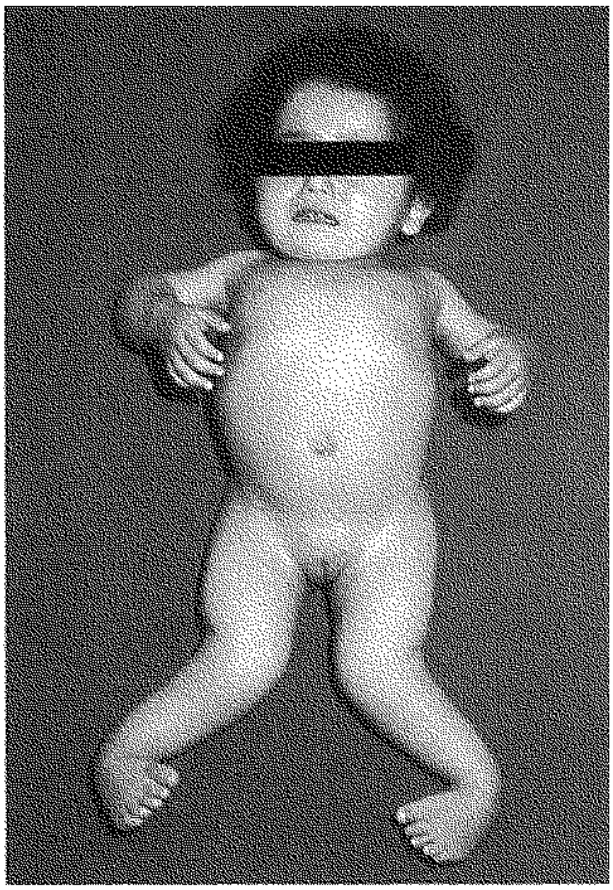

*Fig. 1.* — Typical apparance of this case with Larsen's syndrome (multiple dislocations, flat face, hand and foot abnormalities).

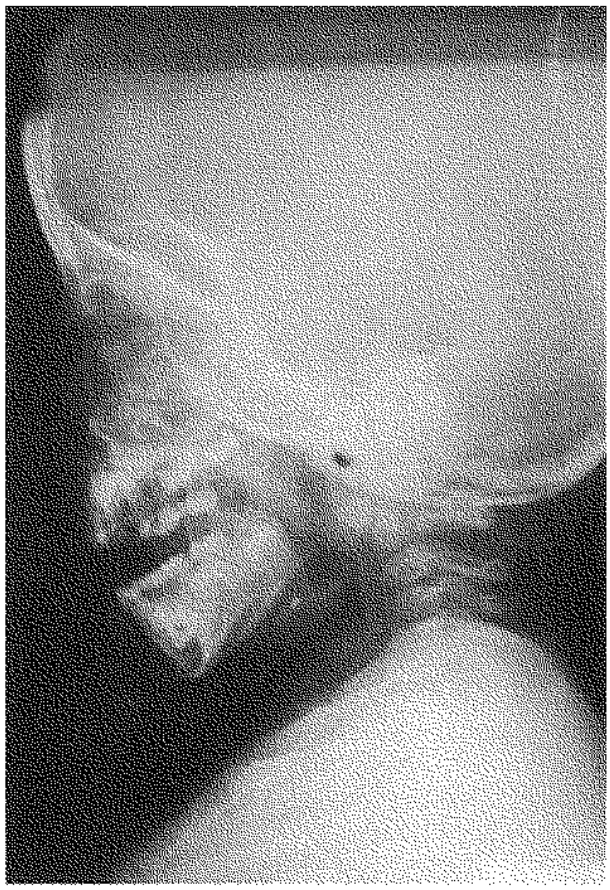

*Fig. 2.* — Lateral cervical radiograph (subluxation between C3 and C4).

rhonchi on pulmonary auscultation. These were thought to be caused by infection. After treatment of the infection, the rales and rhonchi disappeared, but stridor did not. After treatment, the expiration was also normal and silent.

Orthopedic and radiological examination of the patient showed multiple deformities : bilateral dislocation of hip, knee and elbow, barrel chest deformity due to kyphosis and mild thoracic scoliosis, short humeri and bilateral rigid pes equinovarus (fig. 1). There was a 45° valgus deformity of the knees, and the range of motion of the knees was limited to 45-50°. In addition, subluxation between the third and fourth cervical vertebrae (fig. 2), mild thoracic scoliosis (Cobb angle was 22°) and minimal kyphosis were detected. In the hand, radiographs showed short metacarpals and hypoplastic terminal phalanges ; the epiphysis of distal phalanges was not visible. Both tibias were dislocated anterolaterally. There were juxtacalcaneal accessory bones. The talocalcaneal angles were 0°. Computerized tomography (CT) showed that the canal of the cervical vertebrae and the tracheal canal were intact, and there were no internal organ anomalies. The tracheal canal was open in all of the CT sections.

Clinical and otorhinolaryngological examination, tympanogram, measuring of BAEP (Brainstem Auditory Evoked Potential) and laryngeal ultrasonogram showed laryngomalacia and a mixed-type hearing loss on the right side. Otoscopic examination revealed normal tympanic membranes. On measuring BAEP (Brainstem Auditory Evoked Potential) with 90 dB, the latency from I to V for

*Fig. 3.* — Laryngeal ultrasonogram (widening on expiration and semicollapse of larynx on inspiration).

the left ear was 1.76 msec and 5.88 msec, respectively, but no meaningful response was noted in the right ear. The tympanogram confirmed the results of the BAEP.

The laryngeal ultrasonogram (US) showed semicollapse of the larynx on inspiration (fig. 3). On the other hand, there was no tracheal collapse in the cervical trachea.

No numerical or structural abnormalities were noted on the chromosomal analysis which was performed with the method of trypsin–Giemsa–G banding.

Surgical treatment was elected to correct the bilateral hip dislocation. We started by performing a right iliac osteotomy and open hip reduction. During the anesthetic procedure, induction was maintained with sevoflurane via a face mask and after intravenous cannulation, muscle relaxant was given and endotracheal intubation was performed using an endotracheal tube (ETT) with 4-5 mm internal diameter (id) and 15-16 centimeter long and uncuffed. During the surgical procedure, the anesthesiologist had difficulties managing the cardiorespiratory status of the patient.

At one point, events became quite chaotic, and although the critical cardiorespiratory situation was partially due to the displacement of the ETT from its original position, it is our opinion, in retrospect, that part of the situation was due to this young patient's laryngomalacia. At the end of the operation, a hip spica cast was made, and the patient was awakened without any further problems.

## DISCUSSION

This child with multiple joint dislocations had a typical Larsen's syndrome with the characteristic flat face, hand and foot anomalies and other typical findings. Furthermore, there was a juxta-calcaneal accessory bone or two calcaneal ossification centers, which according to Latta at al. may be specific for this entity (9, 11). The case was probably sporadic, because there were no similar cases in the family in spite of the mother and father being first-degree relatives. The case showed rare findings such as mixed-type hearing loss and laryngomalacia, which were reported in a few previous papers. We found only two cases of Larsen's syndrome with mixed-type hearing loss reported since 1990 (15, 18). Moreover, isolated laryngomalacia in Larsen's syndrome has not been previously reported. The laryngomalacia in our patient was not associated with tracheomalacia. The latter is a specific lesion that produces expiratory stridor of a wheezing nature secondary to collapse of the trachea on expiration (14), whereas expiration in our patient was quite comfortable and without any stridor. Moreover, the diagnosis of isolated laryngomalacia was confirmed by CT and by US. Some previous reports have mentioned that patients with Larsen's syndrome may present various fatal risks such as spinal instability, tracheomalacia, heart disease and severe respiratory infection (11). Micheli *et al.* reported that a child with Larsen's syndrome died because of a probable spinal cord injury due to cervical spine instability (12). In the present report, following careful evaluation, prophylactic bracing was used, and early surgical stabilization was considered. The pulmonary infection of the child was treated before the operation.

This case shows that one must be very cautious when operating on such children who have isolated laryngomalacia. The region of laryngeal anomaly may collapse during the operation, resulting in insufficient oxygen intake due to blocking of the mouth of the endotracheal tube. In addition, mobile infolding arytenoid cartilage has been reported in

such children, but we could not document this in our patient (7). We suggest that this problem may be overcome by careful evaluation and by using a longer endotracheal tube for tracheal intubation.

To conclude, the orthopedic surgeon must evaluate the patient with Larsen's syndrome with caution and must be aware of the potentially fatal risks stated above in order to avoid dangerous situations during the operation.

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## SAMENVATTING

*K. KARAKAS, E. F. PERCIN, S. PERCIN. Heelkundige risicofactoren bij het syndroom van Larsen.*

De auteurs bespreken het geval van een meisje met typisch syndroom van Larsen. Naast doofheid van het gemengde type waren er meerdere heelkundige risicofactoren, zoals laryngomalacie en vervicale instabiliteit. De literatuur beschrijft enkele gevallen van Larsen syndroom waarbij risicofactoren zoals cervicale instabiliteit hebben geleid tot een fatale afloop bij ingreep. Orthopaedische chirurgen, die een ingreep plannen bij dergelijke patiënten, hebben er alle belang bij op de hoogte te zijn van laryngomalacie en andere risicofactoren, aanwezig bij het patiëntje in kwestie.

## RÉSUMÉ

*K. KARAKAS, E. F. PERCIN, S. PERCIN. Facteurs de risque chirurgicaux dans le syndrome de Larsen.*

Les auteurs rapportent le cas d'une enfant de 32 mois qui présentait un syndrome de Larsen typique. Elle avait une surdité de type mixte et présentait plusieurs facteurs de risque pour la chirurgie, en particulier une laryngomalacie et une instabilité cervicale. On trouve rapportés dans la littérature quelques cas de syndrome de Larsen où des facteurs de risque tels qu'une instabilité cervicale ont entrainé une complication fatale à l'occasion d'une opération. La laryngomalacie et les autres facteurs de risques retrouvés chez notre patiente sont importants à connaître pour les orthopédistes, en prévision d'un traitement chirurgical.