Introduction: In the present study we report 36 cases of anterior encephaloceles treated at Clinic of Neurosurgery in the University Clinical Center of Kosova over a 24 year period. Materials and methods: All 36 children were included in this retrospective study (1986 through 2009). Their ages ranged from 1 day to 10 years (mean 13 months); 20 were boys and 16 were girls. The commonest type of anomaly seen was nasofrontal 17 patients, 12 nasoethmoidal, and 7 nasoorbital. The size of the lesion varied from 2.5 cm to 28 cm. Hypertelorism occurred in 12 patients. Hydrocephalus was present in 11 patients and in 8 of them was progressive. Results: 1 patient died on 10th postoperative day due to fulminant meningitis. Cosmetic results were judged from parents as excellent in 16 patients, good in 12 patients, average in 6 patients and poor in 2 patients. None of patients were lost to follow – up. Discussion: Histologic examination of the herniated tissue can vary between normal brain to fibrous atrophic nonviable tissue. No familial cases have been reported in the literature, we also have similar experience. In our study we have found that the prevalence is higher among patients coming from rural parts of our country with pore living conditions, malnutrition and so on. We have avoided surgery in very young children until body weight is around 5-6 kg because of complication from blood lose and hypothermia. Conclusion: The aim of treatment is early removal of the meningoencephalocele to allow normal growth forces to be re-established. In patients with hypertelorism, correction surgery is done in the same session. If hydrocephalus is not treated before corrective surgery for encephalocele, the risk of postoperative CSF is very high. With one stage surgery excellent results can be achieved. Keywords: Anterior encephaloceles, craniofacial malformations, pediatric neurosurgery, sincipital encephalocele.

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=0.44, P>0.05), (table 1). For testing of the data we have used X²-test with significant level P<0.05 or P<0.01.

2.2. Radiology

CT scans were obtained in all cases. In 27 of them it was performed 3D reconstruction of the skull. Hydrocephalus was present in 11 patients in 7 of them cerebral index was less than 2.2cm. 2 patients had corpus callosum agenesis.

2.3. Operative procedure

One patient was precluded from surgery because of severe mental retardation. 35 patients underwent surgical reconstruction, 8 patients underwent ventriculi peritoneal shunt medium pressure before corrective surgery. 5 cases underwent extra cranial repair alone. Trans cranial approach was done in 31 patient. Surgical incisions include a bicoronal scalp flap, giving wide exposure of the craniofacial skeleton. Bilateral frontal craniotomy was performed where bone was removed as a free graft. After the elevation of frontal lobe the herniated brain was clearly seen. The dura was opened and the cerebral herniation inspected. Herniated brain in majority of cases consists of gliotic non-functional neural tissue and was aspirated. Bone defect was closed with bone graft that was taken from cranium. In 12 patients osteotomies are performed to translocate the orbit medially.

3. RESULTS

The commonest type of anomaly seen was nasofrontal 17 patients, 12 nasoethmoidal, and 7 nasoorbital, with no significant difference by localization (X²-test =4.167, P>0.05). The size of the lesion varied from 2.5cm to 28 cm. In 9 patients the contents were cystic in 7 of them when baby cried pulsation was felt over the swelling, 27 cases the contents were a mixture of solid and cystic mass with significant difference by content (X²-test =9, P<0.01) (Table 2). Hyperelorism occurred in 12 patients. Hydrocephalus was present in 11 patients and in 8 of them was progressive. In 32 patients the skin coverage was complete and in 4 cases thin layer of epidermis was covering the lesion. In 1 case the lesion was extremely large and trampled on the nose and eyes, causing severe difficulties with breathing and vision because of this required urgent operative treatment. The average paternal age was 28.2 years. Consanguinity was reported in 1 family. There was no family history of other encephaloceles or any neural tube defect. In 3 cases epileptic seizures were present. From 36 children 3 of them were going to school before the surgery and 2 of them reported teasing from other children. Other congenital anomalies like cleft palate, cleft lip, absence of nostril, microphthalmia, syndactyly were noted in 1 patient each.

From all number patients in our study, which included 36 patients, 34 patients, who underwent correction, survived the operation. Immediate postoperative CSF leak through the skin was observed in 1 patient which required tighter skin closure and a tight head bandage. In 1 patient bone flap osteomyelitis was observed she underwent repeated surgery and received antibiotic therapy until the infection healed completely the cause was: Pseudomonas aeruginosa. 1 patient died on 10th postoperative day due to fulminant meningitis. In spite of anticonvulsant therapy 1 children continue to seize even after 8 years following surgery. Nasolacrimal dysfunction usually persisted postoperatively. Cosmetic results were judged from parents as excellent in 16 patients, good in 12 patients, average in 6 patients and poor in 2 patients. None of patients were lost to follow – up.

4. DISCUSSION

Anterior encephaloceles are common in many southeast Asian countries such as Burma (19, 23), Thailand (14,
18), Cambodia (11), Malaysia (13), and Indonesia (12). In Thailand, it has been shown that frontoethmoidal MECs occur in one of 5000 to 6000 live births (14, 18). A few authors have described these deformities in Australian aboriginal people (13) and in some patients in African countries (13), and in India and Pakistan (9, 10). This lesion are rare in North America, Japan and Wester Europe. Till date, the exact etiology is not known, although various hypotheses have been proposed (5, 6). Currently, two schools of thought exist regarding the origin of these lesions. The first concept involves the point of weakness in the facial skeleton. The frontal bone is a membranous bone that forms from the underlying dura, whereas the ethmoid bone develops from endochondral bone formation. The function of the membranous and endochondral bone (foramen cecum is thought to result in a week area through which the neural elements can herniate (2, 7). A second hypothesis states that a delayed closure of the neural tube ultimately prevents normal union of the facial bone (18). Findings of a trapped meningocele and peripheral nerve elements, as well as isolated neural tissue remnants along the original tract appear to substantiate this second theory. However, the fact that most anterior encephaloceles are covered with normal skin indicates that these defects are not simply a failure of neurulation. Thus, the precise etiology of these lesions remains speculative. Histologic examination of the herniated tissue can vary between normal brain to fibrous atrophic nonviable tissue (4). No familial cases have been reported in the literature, we also have similar experience. Suwanwela (16) described a pair of identical twins where 1 twin was not affected, concluding that a genetic mechanism was not the primary cause. Other theories have included a simple vitamin deficiency however, Kyu and Thu (20), in a large series in Burma, revealed that over half of the mothers of (16); affected children were on vitamins during their pregnancy. In our study we have found that the prevalence is higher among patients coming from rural parts of our country with poor living conditions, malnutrition and so on. We have avoided surgery in very young children until body weight is around 5-6 kg because of complication from blood lose and hypothermia.

5. CONCLUSIONS

In a country such as Kosova, the treatment of specific disease processes must be adapted to fit current local conditions. The development of low-cost procedures is very important. Three-dimensional CT scan has been a major improvement in investigation of defects in craniofacial skeleton. The aim of treatment is early removal of the meningoencephalocele to allow normal growth forces to be re-established. In patients with hypertelorism, correction surgery is done in the same session. If hydrocephalus is not treated before corrective surgery for encephalocele, the risk of postoperative CSF is very high. With one stage surgery excellent results can be achieved.

REFERENCES