FACIAL CLEFT AND MAXILLARY DUPLICATION

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SUMMARY

Facial clefts and facial duplications are very rare congenital anomalies. We present two cases. The first is a case of maxilla and upper lip duplication with lateral cleft lip. The second case is a maxilla duplication with a Tessier No: 30 mandibular cleft. We compared and discussed our cases with rare similar anomalies found in the literature.

Key Words: Facial cleft, Maxillary duplication

INTRODUCTION

Facial clefts and facial duplications are rare occurrences. These anomalies may occur together or singly. Isolated facial clefts are seen more frequently than facial duplications and facial duplications with facial clefts (1-3).

In the literature, a small number of facial duplications and a larger number of facial clefts have been reported (2-10).

Etiology of both facial clefts and facial duplications are unknown and also morphopathogenesis is unclear (1,2).

CASE REPORTS

CASE 1: A male newborn baby was admitted as an emergency case to the plastic surgery unit. He was the first baby of normal and healthy parents. The mother was 20 and the father was 22 years old. Pregnancy and delivery were normal at full term. There was no family history of craniofacial anomalies.

Physical examination showed an intraoral mass of 3.5 x 3 x 2 cm which separated the maxilla and mandible from each other and prevented closure of the mouth (Figs 1a, 1b). The mass caused airway obstruction so emergency surgery was required. In addition to the mass the baby had a left lateral cleft lip, a 1.5 x 2 cm capillary hemangioma on the scalp, a left choanal atresia and a pectus excavatus deformity. The mass originated from the left side of the palate and left buccal mucosa and was supplied by a branch of left greater palatine artery. The mass was totally excised (Fig 2), leaving a large raw area in the left side of the hard palate (Fig 3). After the excision of the mass the raw surface on the palate was left open for secondary mucosal epithelization.

Macroscopically the mass was composed of two parts. Posterior aspect had a hard core covered with mucosa and anterior aspect had a soft core which was covered with skin. Histopathological examination showed that hard core contained bony tissue and was covered by pseudostratyphied columnar epithelium (Fig 4). The superior side of the soft part of the mass was covered by a strip of mucosa similiar to vermillion containing sebaceus glands.

The postoperative progress was uneventful (Fig 5). The lateral cleft lip (Fig 5) was repaired by the Skoog technique (Fig 6). The left choanal atresia was corrected at the age of 6 months. A cleft palate repair is planned at the age of 18 months.

CASE 2: A male newborn baby with normal, healthy parents (the mother and father were both 20 years old) was admitted to plastic surgery department at the sixth hour after the delivery. Physical examination showed a mongoloid baby with hypertelorism, a bifid nose with a short columella and a mass covered with mucosa on the hard palate (Figs 7a, 7b). There was no family history of craniofacial anomalies. Pregnancy and delivery were normal. An X - ray showed a median cleft of the mandible (Tessier No: 30 cleft) (Fig 8) and a spina bifida occulta in the lumbar region (Fig 9).

Operation was planned but cardiopulmonary arrest caused death of the baby two hours after the admisson. Parents did not give permission for autopsy.

DISCUSSION

Reports of facial duplication with facial clefts are very rare. Isolated facial cleft cases are more frequent than facial duplications. Facial duplication cases without facial clefts are very rare and usually maxilla and mandible duplication are seen together, which are reported as 'accessory mouth' (11). Early reports include the cases of Mc Laughlin (10), Beatty (7) and Borçbakan (11). Maxilla and mandible duplications are also very rare and almost all of these cases are seen with rare facial clefts.

In Fearon and Mulliken's study a child with dublicate maxilla was presented along with the review of the literature of similar cases (2). Fearon and Mulliken's case had similarities and differences with ours. The intraoral mass of our first case resembled Fearon and Mulliken's case. The intraoral masses in Fearon and Mulliken's study and masses in our cases were similar in size and composition.

Additionally, both Fearon and Mulliken's case and our two cases had palate defects (cleft palate). But Fearon and Mulliken's case was different in having a left unilateral cleft lip and left hypoplastic eye and orbit, whereas our first case was accompanied with lateral cleft lip (Fig 5).

The maxillary duplication of our second case resembled Bhattacharya et al. 's and Chowdhury and Roy's cases (4,5). Since, our second case died before surgery, we could not discuss the detailed characteristics of the mass. Bhattacharya's case had a Tessier No: 3 + 4 + 7 cleft, our case had Tessier No: 30 cleft. Our case additionally had orbital hypertelorism and spina bifida occulta (Figs 8,9), which were not present in the before mentioned reports.

Bhattacharya et al. reported his case of alveolar arch duplication with a Tessier No: 3 + 4 + 7 cleft and claimed his case as the first case of duplication in the literature. However, similar cases have been reported by Fearon and Mulliken (2), Goulian and Conway (6), Borçbakan (11). But these authors did not name the pathology as maxillary or mandible duplication perhaps because it was difficult to classify these cases in groups as the etiology was unknown and the morphogenesis was unclear.

Terminology such as "parasitic fetus", "teratoma" or "accessory mouth" were used in the past for these cases. Today these terms are not used. In the differential diagnosis the widely accepted theory claims that multipotential early embryonal cells cause duplication (2).

The main problem with the rare cases is classification. We believe that this problem will be solved with the reports of further cases and with further studies regarding the etiology and morphogenesis.

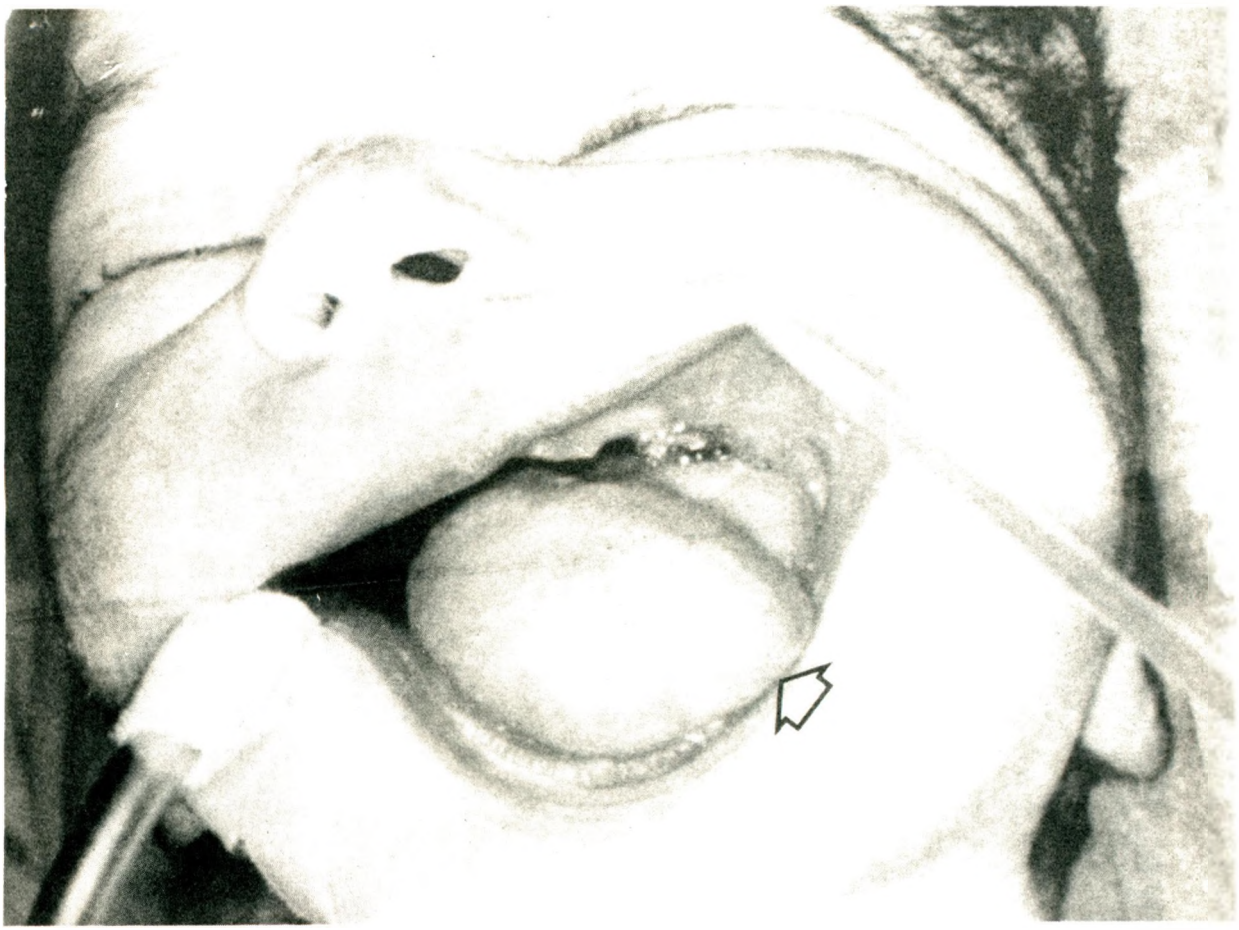

Fig 1 a- Maxillary duplication (black arrow). Preoperative appearance of the 1st case.

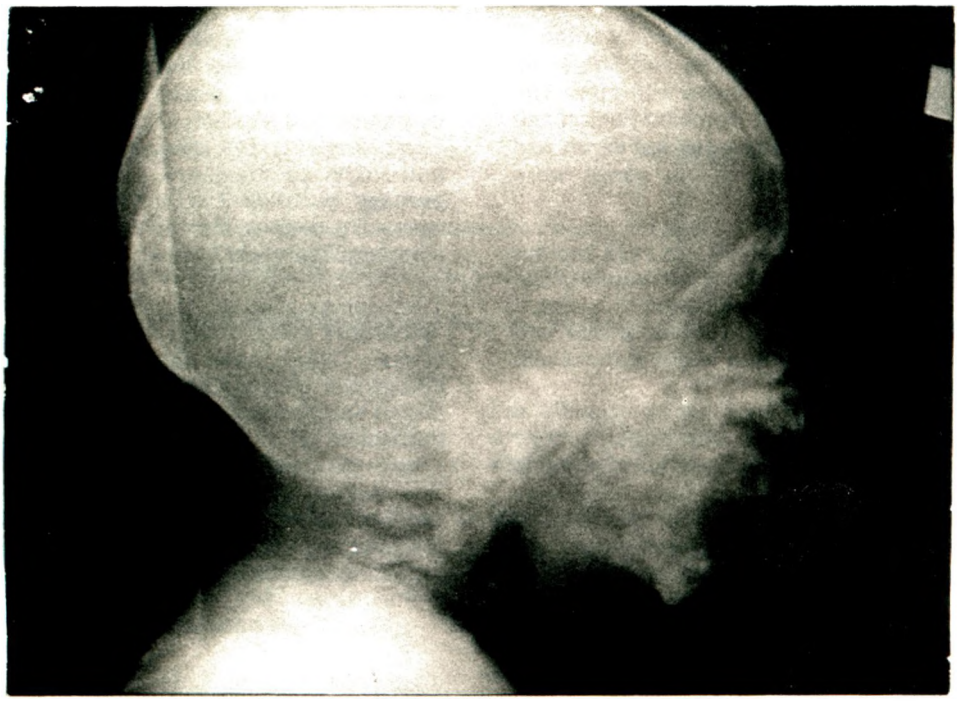

Fig 1 b- Maxillary duplication (black arrow) on preoperative lateral skull roentgenogram of the 1st case.

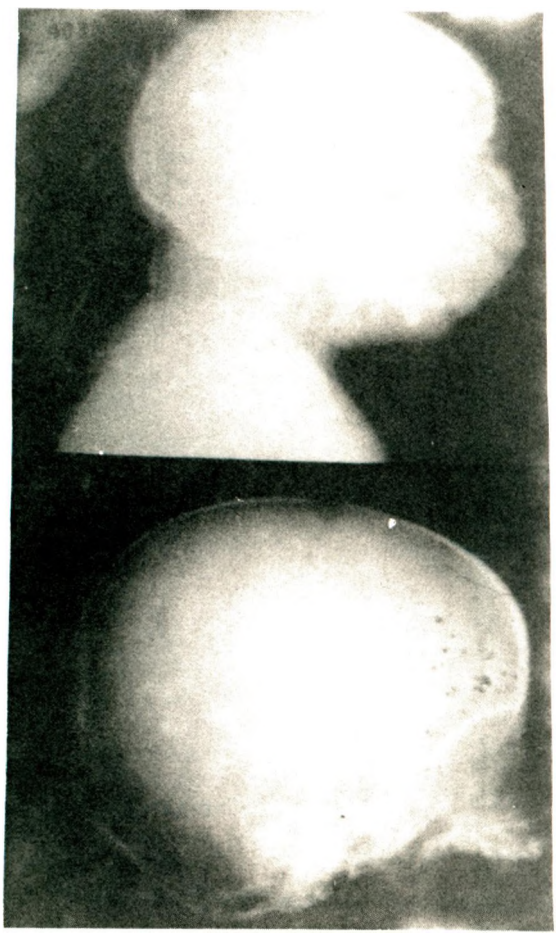

Fig 2- (a) Preoperative lateral skull roentgenogram of the 1st case.
(b)Postoperative lateral skull roentgenogram of the 1st case.

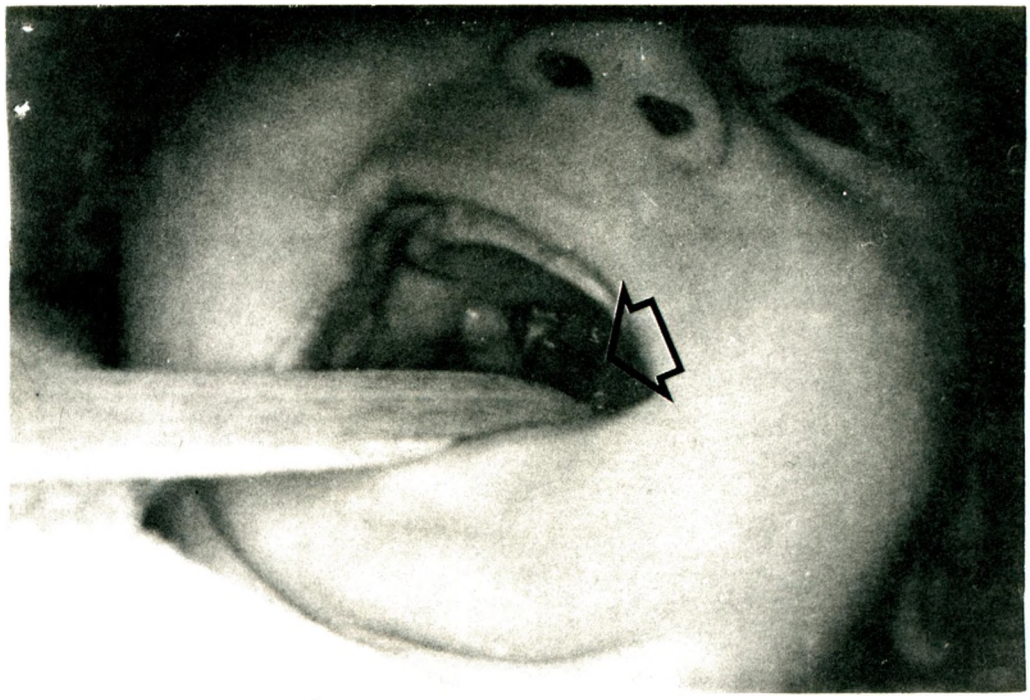

Fig 3- Cleft palate (black arrow) of the 1st case.

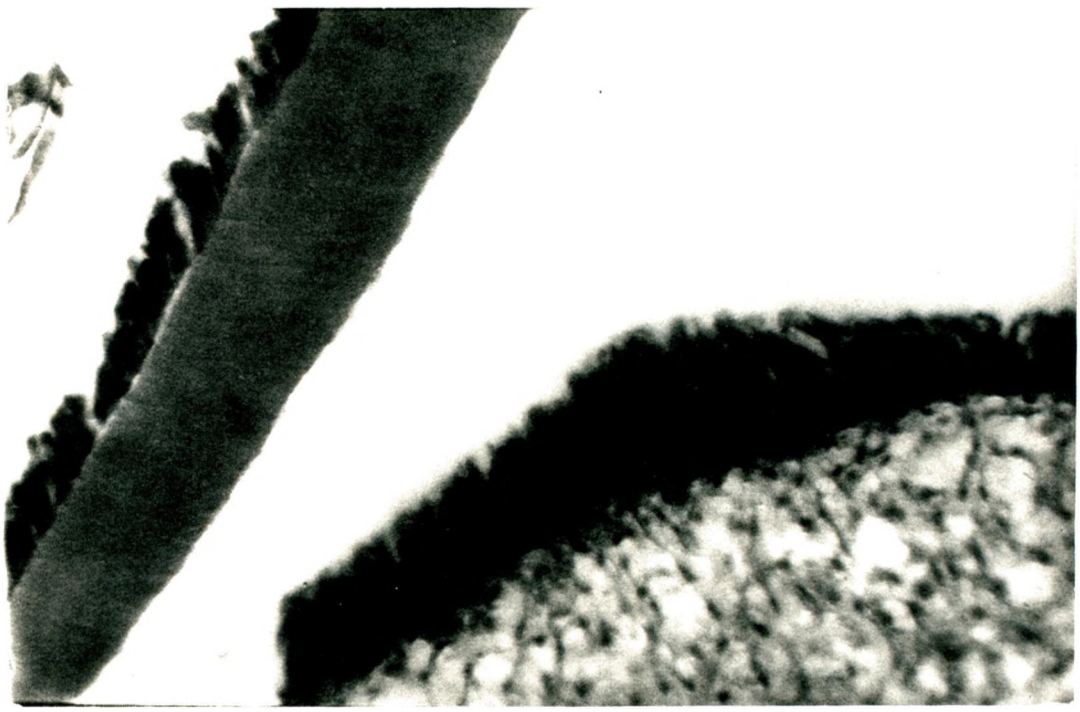

Fig 4- Pseudostratyphied columnar epithelium (X100)
The section is of mucosal lining from the posterior segment of the mass.

Fig 5- Early postoperative appearance of the 1st case. Black arrow-lateral cleft lip.

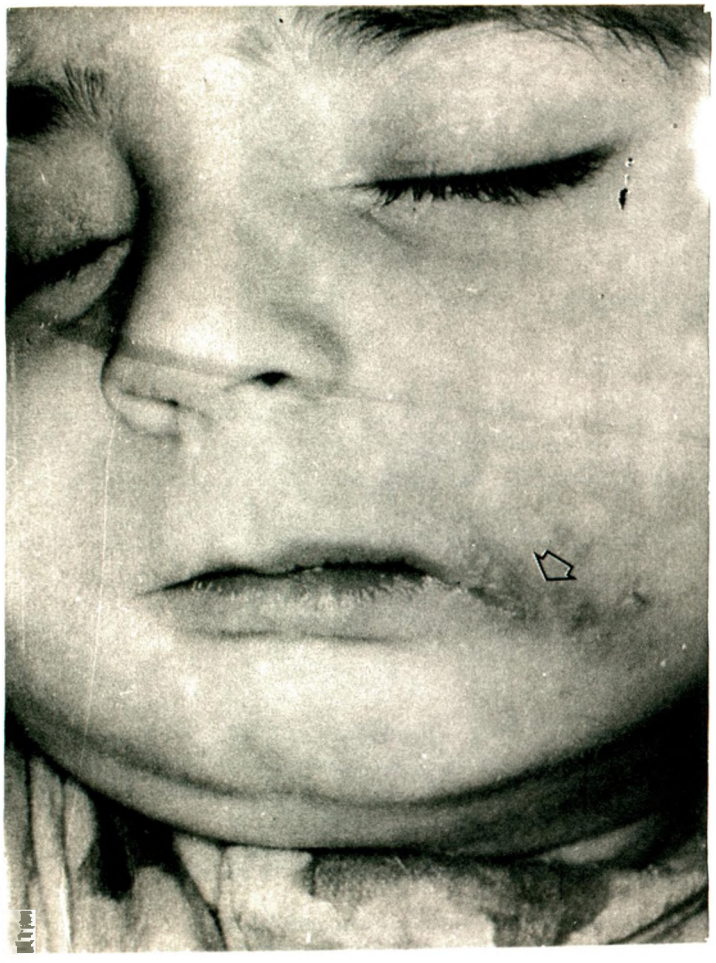

Fig 6- Late postoperative appearance of the 1st case following lateral cleft lip repair (10 days later)

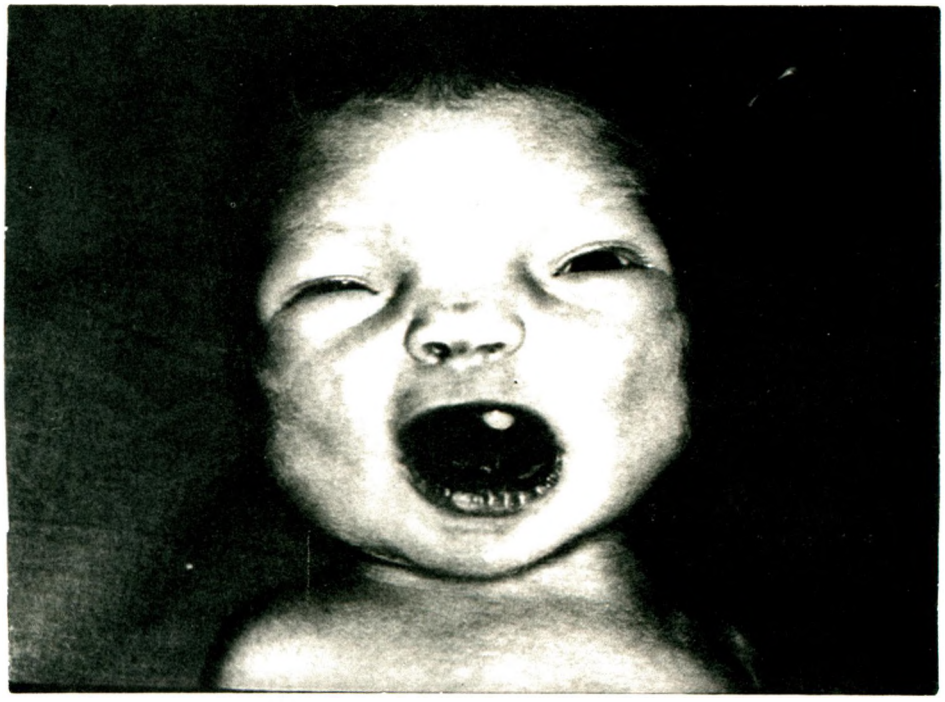

Fig 7 a- Preoperative appearance of the 2nd case

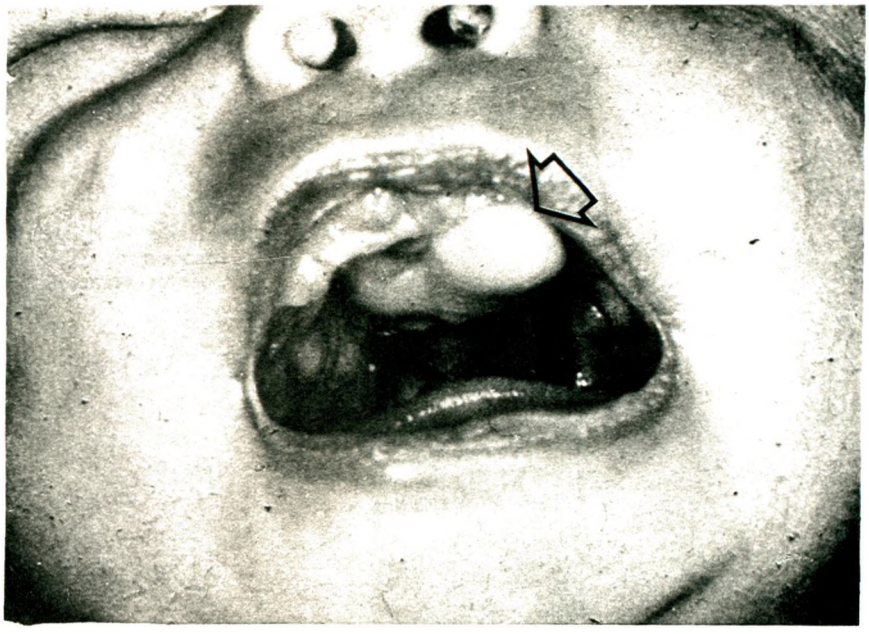

Fig 7 b- Maxillary duplication (black arrow). Open mouth view of the 2nd case.

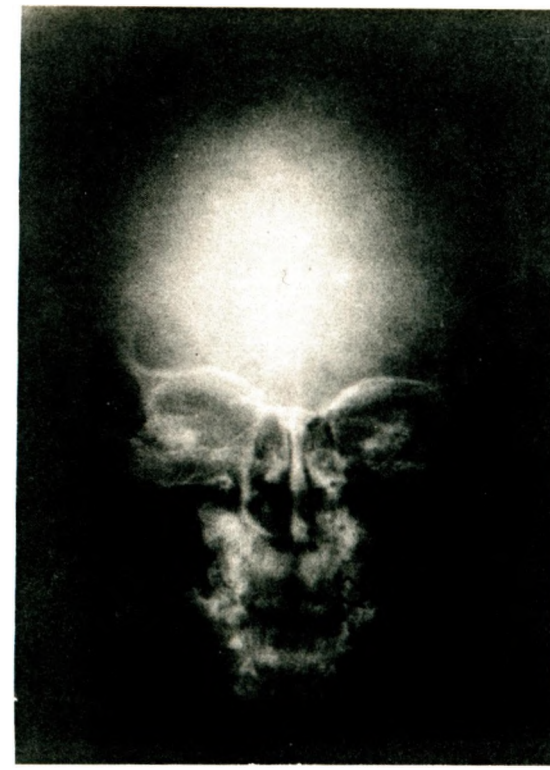

Fig 8- Maxillary duplication (black arrow). Skull roentgenogram of the 2nd case.

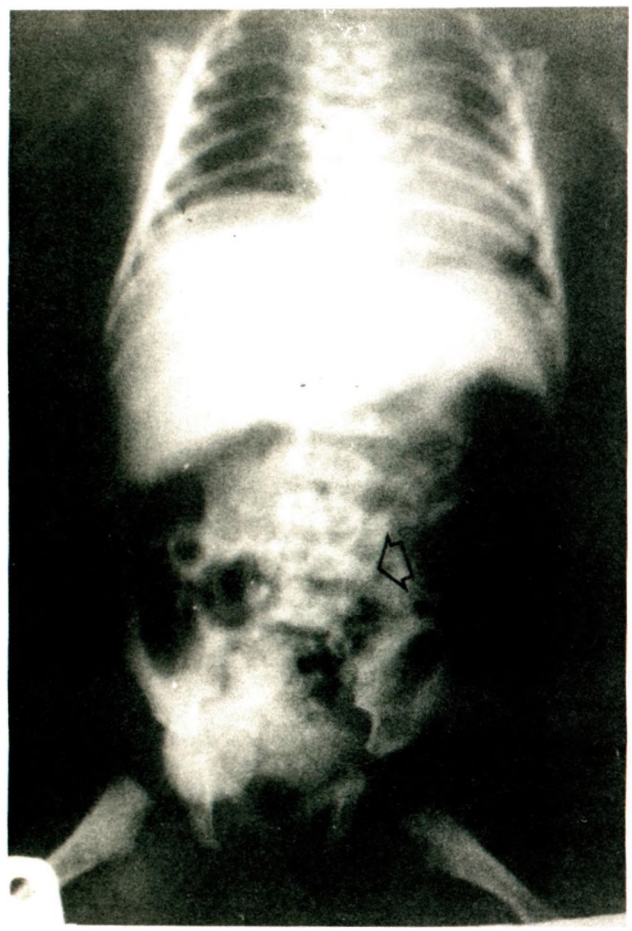

Fig 9- Lumbar spina bifida occulta (black arrow). Total body roentgenogram of the 2nd case.

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Özellikleri • Endikasyonlar

Prolaktin memede bazı fonksiyonlarını önemli ölçüde kontrol etmektedir. Prolaktin inhibitörü Dopergin, bundan dolayı normal süt produksiyonu ve salgılanmasını kesmek (primer ve sekonder süten kesme) ve meme bezinin patolojik durumlarını (galaktostaz, mastitis, galaktore) gidermek için uygundur. Mastodini ve diğer premenstrüel rahatsızlıklar da, Dopergin ile yapılan bir tedaviye iyi cevap verirler.

Yüksek bir prolaktin düzeyi gonad fonksiyonlarını inhibe eder. Bundan dolayı yüksek bir prolaktin düzeyinde görülen amenore ve diğer siklus bozukluklar ve bunlara bağlı olarak oluşan infertilite, Dopergin ile tedavi edilebilir. Bu semptomların erkekteki karşılığı libido ve potens bozukluklarıdır.

Yukarıda bildirilen rahatsızlıkların nedeni prolaktin salgılayan hipofiz tümörleri olabilir. Makroadenomlarda Dopergin uygulaması sonucunda klinik semptomların düzelmesinin yanısıra, sellada tümor boyutları da belirgin biçimde küçülür.

Akromegali'li hastaların bir kısmında, büyüme hormonunun aşırı produksiyonu olumlu yönde etkilenmektedir. Dolayısıyla bu gibi durumlarda diğer tedavi biçimleri yetersiz kalındığında ya da mümkün olmadığında, Dopergin kullanılabilir.

Endikasyonlar -Prolaktin inhibitörü olarak: Primer ve sekonder süten kesme (tıbbi açıdan endike ise), mastitis, galaktore, postpartal süt konjestiyonunun azaltılması; yüksek prolaktin düzeylerine bağlı amenore ve diğer siklus bozukluklar (örn. sarı cisim yetmezliği); kadında yüksek prolaktin düzeylerine bağlı infertilite ve erkekte libido ve potens bozuklukları (örn. hipofiz tümörü sonucu); yüksek prolaktin düzeylerine bağlı premenstrüel şikayetler (örn. mastodini).

Dozaj ve uygulama için ön saylaya bakınız. Tabletler her zaman yemek esnasında bir miktar gıda ile birlikte alınmalıdır. İlacı karşı daha iyi tolerans, genellikle dozu yavaş yavaş arttırarak sağlanır.

Yan etkiler Genellikle tedavi başlangıcında, dozun hızla yükseltildiği veya yüksek doz uygulandığı durumlarda, mide bulantısı, baş ağrısı, baş dönmesi, yorgunluk hissi, terleme ve seyrek olarak kusma görülebilir.

Tek tük olgularda, özellikle individuel hassasiyeti olan kişilerde ani kan basıncı düşmeleri (ortostatik kollaps'a neden olabilecek) ve şiddetli kusma nöbetleri gözlenmiştir. Bu gibi durumlarda Sulpirid (100 mg a kadar) i.m. uygulanmalıdır. (Dikkat: Şiddetlenen Parkinson semptomatiği mümkündür.)

Bu tür belirtiler genelde tedavinin yanında kesilmesini gerektirmez ve doz azaltılması yoluyla kontrol edilebilir. Muhtemelen tedavide, doz önemli derecede yükseltilse bile, bu yan etkiler ortaya çıkmamaktadır.

Kontrandikasyonlar Periferde ve kalpte (koroner yetmezliği) ağır arteriyel dolaşım bozuklukları olan hastalarda endikasyon özenle tartılmalıdır.

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