Case Report / *Olgu Sunumu*

Doi: 10.4274/npa.y6311

Cerebellar Mutism Syndrome After Posterior Fossa Surgery: A Report of Two Cases of Pilocytic Astrocytoma

Posterior Fossa Cerrahisi Sonrası Ortaya Çıkan Serebellar Mutizm Sendromu: Pilositik Astrositoma Tanılı İki Olgunun Sunumu

Hasan Burak GÜNDÜZ[1], Mustafa İlker Kuntay YASSA[2], Ali Ender OFLUOĞLU[1], Lütfü POSTALCI[1], Erhan EMEL[1]

[1]Bakırköy Prof. Dr. Mazhar Osman Mental Health and Neurology Training and Research Hospital, Clinic of 2nd Neurosurgery, İstanbul, Turkey
[2]Bakırköy Prof. Dr. Mazhar Osman Mental Health and Neurology Training and Research Hospital, Clinic of 1st Neurosurgery, İstanbul, Turkey

ABSTRACT

Cerebellar mutism is a type of syndrome including decreased speech, hypotonia, ataxia and emotional instability which occurs after posterior fossa surgery. It has been first reported by Rekate et al. and Yonemasu in 1985. It is well known that long tract signs and lower cranial nerve involvement are not seen with this syndrome and understanding is preserved. However, the pathophysiology of cerebellar mutism has not been well clarified yet. It is mainly seen in patients with medulloblastoma and brainstem involvement. In this report, we present two extraordinary cases of cerebellar mutism after posterior fossa surgery. They were considered extraordinary because their hystopathological analysis results yielded pilocytic astrocytoma which is out of the predefined risk factors. *(Archives of Neuropsychiatry 2013; 50: 368-371)*

Key words: Cterebellar mutism, pilocytic astrocytoma, posterior fossa surgery

Conflict of interest: The authors reported no conflict of interest related to this article.

ÖZET

Serebellar mutizm sendromu posterior fossa cerrahisi sonrası azalmış konuşma, hipotoni, ataksi ve duygusal kararsızlık ile kendini gösteren bir sendromdur. İlk kez 1985 yılında ayrı makalelerde Rekate ile arkadaşları ve Yonemasu tarafından rapor edilmişlerdir. Anlamanın korunmuş olmasının, uzun trakt bulgularının ve alt kranyal sinir tutulumunun bu sendromda görülmediğinin iyi bilinmesine rağmen, serebellar mutizmin patofizyolojisi henüz ayrıntılı olarak açığa çıkarılamamıştır. Sıklıkla medulloblastomalı ve beyin sapı tutulumu olan hastalarda görülmektedir. Posterior fossa cerrahisi uyguladığımız iki olgumuzda serebellar mutizm sendromu gelişmiştir. Kabul edilen risk faktörlerinin dışında, histopatolojik inceleme sonucunun pilositik astrositoma gelmesi nedeniyle sıra dışı olarak kabul ettiğimiz bu iki olguyu sunmaktayız. *(Nöropsikiyatri Arşivi 2013; 50: 368-371)*

Anahtar kelimeler: Serebellar mutizm, pilositik astrositoma, posterior fossa cerrahisi

Çıkar çatışması: Yazarlar bu makale ile ilgili olarak herhangi bir çıkar çatışması bildirmemişlerdir.

Introduction

Cerebellar mutism syndrome is a well-defined syndrome which is observed rarely after wide removal of posterior fossa tumors (1). It is characterized with decreased speech or loss of speech, hypotonia, ataxia and emotional instability (2,3).

It was reported by Rekate et al. and Yonemasu in different articles in 1985 for the first time after posterior fossa surgery (1,4,5). More than 200 cases of cerebellar mutism syndrome have been reported until the present time. Its incidence is 11%-29% (6). It is frequently observed in patients with medulloblastoma and brainstem involvement (6,7,8,9,10,11,12). According to Gudrunardottir's literature review, definite findings in the area of astrocytoma and ependimoma are insufficient, although the incidence of cerebellar mutism in children with medulloblastoma has been documented well (6). In this article, we present two patients (one male adolescent and one female young adult) who were diagnosed with pilocytic astrocytoma and developed cerebellar mutism after posterior fossa surgery.

Case 1

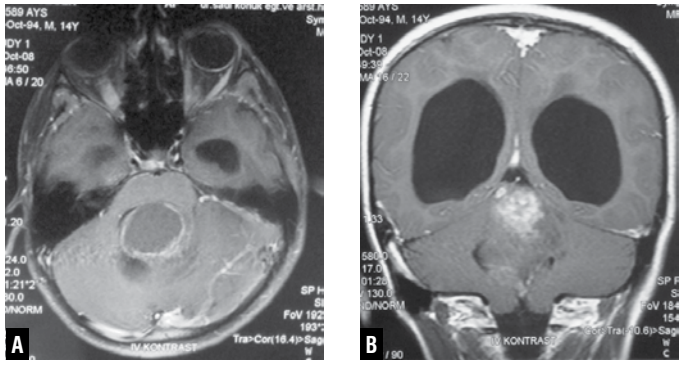
A 14-year old right-handed male patient presented to our clinic with complaints including headache, speech disorder,

Correspondence Address/Yazışma Adresi
Hasan Burak Gündüz MD, Bakırköy Prof. Dr. Mazhar Osman Mental Health and Neurology Training and Research Hospital, Clinic of 2nd Neurosurgery, İstanbul, Turkey
Gsm: +90 532 573 31 20 E-mail: bgunduz62@yahoo.com **Received/Geliş tarihi:** 16.09.2011 **Accepted/Kabul tarihi:** 27.06.2012

nausea and vomiting and balance disorder. On neurological examination, ataxia, dysmetria and dysdiadochokinesis were found. On magnetic resonance imaging, a mass with a size of 3x4x4 cm which also had a cystic component was observed in the posterior part of the 4th ventricle (vermian location). In addition, hydrocephalus was also present (Picture 1). The patient was started to be operated in the sitting position. The tumor was removed almost completely by vermian incision using surgery microscope (Picture 2). The pathological diagnosis was pilocytic astrocytoma. After the operation decreased speech was observed in the patient. However, his understanding was preserved. External ventricular drainage was started because of hydrocephalus. On the third post-operative day, the patient was aphasic. His movements were markedly slowed down. At the end of the second month, the patient started to speak gradually. However, dysarthria and slowness in speech continued. In the post-operative 3rd year, his problems in speaking were eliminated except for dysarthria. The patient still continues his education, though with delay.

Case 2

A 20-year-old female patient presented to our clinic with complaints including postural disorder, a feeling of dizziness, nausea, vomiting and headache. On neurological examination, she had bilateral nistagmus. His balance in the standing position was disrupted. She had dysmetria in the left side. On cranial magnetic resonance imaging, a tumor which had heterogeneous contrast uptake was found in the posterolateral part of the 4th ventricle in the cerebellum (Picture 3). The patient was started to be operated in the sitting position. The tumor was removed almost completely by suboccipital craniectomy (Picture 4). Postoperatively, the patient's understanding, speech and movements were evaluated to be natural. On the 4th day, the patient's speaking decreased gradually and ceased. During this process, a marked slowing was observed in her movements. Three months later, speaking started gradually. Speech was dysarthric. Although speech improved to a great extent 3 years after the operation, it preserved its dysarthric property. In addition, slowing in the movements and difficulty in writing continued, though they were decreased.



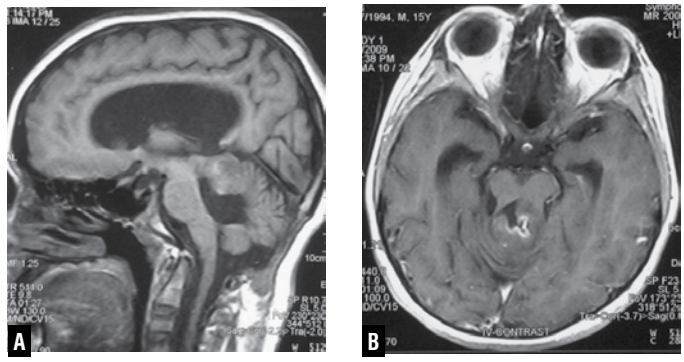
Picture 1. Case1 a) Pre-operative contrast cranial magnetic resonance imaging, T1 sequence, axial section b) Pre-operative contrast cranial magnetic resonance imaging. T1 sequence, coronal section

Discussion

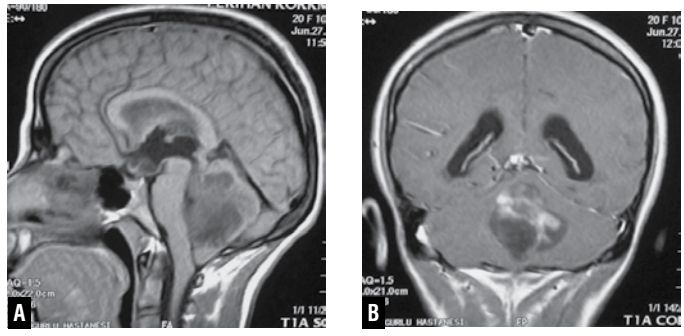
Clinical Findings and Incidence

It is well known that understanding is preserved, long tract findings and lower cranial nerve involvement are not observed in cerebellar mutism syndrome (7). Delayed onset of mutism after operation, its resolution and relation with other neurological findings are also characteristic and well defined.

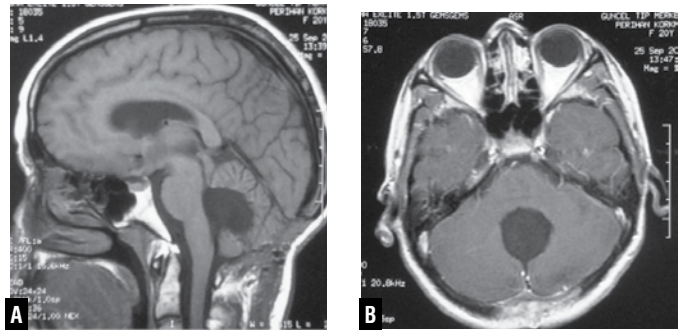
Cerebellar mutism is an expression of mutism state which arises from cerebellar lesions rather than the lower cranial nerves or cerebrum. Its basic characteristics include late onset after surgical intervention (1-6 days), a disease period ranging from 1 day to 4 months and dysarthric speech after recovery (13). The mean time of onset is 1-7 days and the duration is 7-8 weeks (9,14). Recovery is



Picture 2. Case 1 a) Post-operative cranial magnetic resonance imaging, T1 sequence, sagittal section b) Post-operative cranial magnetic resonance imaging, T1 sequence, axial section



Picture 3. Case 2 a) Pre-operative contrast cranial magnetic resonance imaging, T1 sequence, sagittal section b) Pre-operative contrast cranial magnetic resonance imaging, T1 sequence, coronal section



Picture 4. ICase 2 a) Post-operative cranial magnetic resonance imaging, T1 sequence, sagittal section b) Post-operative cranial magnetic resonance imaging, T1 sequence, axial section

spontaneous and rapid and complete in some cases. However, language and speech dysfunctions including ataxic dysarthria, disruption in speech habits and slowed speech continue in most patients in the long term (7,9,10,13,15,16,17,18).

Anatomic localization of the cerebellar lesion and formation theories in mutism preserve their controversial status. Two basic theories which are thought to contribute to formation of cerebellar mutism have been proposed. The first theory is related with the psychological background of cerebellar mutism. Loss of speech in the child may be a psychological reaction to the stress of surgical intervention. "Transient mutism" behavioral changes as stress reaction in the childhood is a well known psychogenic state in which refusal of food is added to mutism.

The second basic theory is related with the organic background of cerebellar mutism. However, the anatomic basis of cerebellar mutism is based on assumptions (1).

Anatomy: Most investigators agree that the basic cause of cerebellar mutism is bilateral interruption of the dentato-thalamo-cortical pathways (12,14,19,20,21,22,23,24,25). The most common explanation of cerebellar mutism emphasizes bilateral damage to the dentate nuclei (7,15,24,26). However, mutism may also occur with damage to any site of the pathway in question. This picture has also been reported in bilateral thalamotomy performed for Parkinmson's disease, in lesions in the supplementary motor area and bilateral edema in the brachium pontis/conjunctivum or superior cerebellar peduncles (8,24,25).

Other studies have also confirmed the importance of involvement of the brainstem (16,17,27). This involvement includes the monoaminergic regions located centrally in the mesencephalon and substantia nigra (6,11,28). In addition, preoperative brainstem compression has been defined as a herald of postoperative mutism (29).

Pathophysiology

The pathophysiological mechanism of cerebellar mutism has not been elucidated fully yet. Although the basis of the event is surgical intervention, the findings do not occur immediately and at least partial recovery is the rule. If mutism starts immediately after surgical intervention, one should be suspicious about bulbar dysfunction which occurs with damage to the cranial nerve nuclei in the brainstem (12). In cerebellar mutism, seconday processes initiated with tumor resection should be considered (19). It is thought that these secondary processes are closely related with perfusional disorders, edema, troubles in release of neurotransmitters and axonal injury (6).

Cerebellar perfusional disorders: during surgical approach to the cerebellum, intraoperative coagulation of the perforating vessels and arterial embolic obstruction may be the cause of cerebellar hypoperfusion, transient ischemia and thus cerebellar mutism.

Edema: Delayed onset of cerebellar mutism may be related with postoperative swelling and edema.

Transient disruption in release of neurotransmitters: According to Siffert et al., changes in the levels of neurotransmitters and disruption of connective structures at synaptic or transsynaptic levels may explain the delay between operation and onset of mutism (6,8,30).

Axonal injury: Surgical intervention directed to the ascendant pathways and strain may be considered important pathological factors (12,14,31). Magnetic resonance imaging and diffusion tensor imaging examinations have shown that functional interruptions of white matter bundles involving the efferent axons in the superior cerebellar pedincules are an important pathophysiological component of cerebellar mutism (23).

Risk Factors

Many risk factors have been defined for cerebellar mutism. One of these is involvement of the brainstem by a tumor (10,12,16). Middle line localization (vermian and 4th ventricle) and tumor type can be added to this. The most common tumor which leads to cerebellar mutism is medulloblastoma in pediatric patients (7,8). Two or 3-fold higher incidence rates have been reported compared to astrocytoma and ependymoma (6,9,21,32). In addition, though not as determinative as the above-mentioned three criteria, brainstem compression, tumor size and preference of vermian incision as the surgical approach are defined as risk factors (6).

Treatment

Medical treatment: The investigators who think that the dopaminergic cell groups in the monoaminergic pathways are important recommend use of bromocriptine to reverse the symptoms of akinetic mutism (11,29). However, this does not necessarily have the desired impact in patients with cerebellar mutism (21). Nimodipine as a calcium antagonist is recommended to prevent ischemia related with vasospasm (33). However, this is not a generally accepted treatment, either (6).

Speech therapy: The level of affection of speech has been used to evaluate and investigate the occurrence of cerebellar mutism. It has been shown that speech monitorization is partially helpful in grading the disease severity and monitoring the process of speech disorder in the post-mutistic phase (23,31). However, there is no concensus about the efficiency of speech therapy in the recovery process other than this (6).

Prevention: Prevention opportunities in cerebellar mutism are still controversial. There are studies suggesting that surgical approaches without separating the vermis (telovelar approach), partial resection in critical regions instead of radical en bloc removal, intraoperative electrophysiological monitorization and avoidance of excessive retraction decrease the risk of mutism (34,35,36,37,38,39).

Cerebellar mutism developed in two patients with posterior fossa tumor who were operated in our clinic. Since the tumors were localized in the middle line and brainstem compression was observed in these patients, they carried the above-mentioned risk factors. The histopathological diagnosis of these two patients was pilocytic astrocytoma.

The incidence of cerebellar mutism has been documented well in children with medulloblastoma. However, it has been reported that there are no sufficient data about patients with astrocytoma and ependimoma in previous literature reviews (6). In addition, the diagnosis of pilocytic astrocytoma is not considered a risk factor for cerebeallar mutism. We published these two cases which did not overlap with the general information to emphasize complications which might develop outside expectations, though we do not have sufficient data to draw a conclusion as a case presentation.

References

1. İldan F, Tuna M, Erman T, Göçer AI, Zeren M, Çetinalp E. The Evaluation and Comparison of Cerebellar Mutism in Children and Adults after Posterior Fossa Surgery: Report of two Adult Cases and Review of the Literature. Acta Neurochir (Wien) 2002; 144:463-473.
2. Wells EM, Khademian ZP, Walsh KS, Vezina G, Sposto R, Keating RF, Packer RJ. Postoperative cerebellar mutism syndrome following treatment of medulloblastoma: Neuroradiographic features and origin. J Neurosurg Pediatrics 2010; 5:329-334.
3. Yıldız O, Kabataş S, Yılmaz C, Altınörs N, Ağaoğlu B. Cerebellar mutism syndrome and its relation to cerebellar cognitive and affective function: Review of the literature. Annals of Indian Academy of Neurology 2010; 13:23-27.
4. Rekate HL, Grubb RL, Aram DM, Hahn JF, Ratcheson RA. Muteness of cerebellar origin. Arch Neurol 1985; 42:697-698.
5. Yonemasu Y. Cerebellar mutism and speech disturbance as a complication of posterior fossa surgery in children. Presented at the 13th Annual Meeting of Japanese Society for Paediatric Neurosurgery, Tsukuba, Japan, 1985.
6. Gudrunardottir T, Sehested A, Juhler M, Schmiegelow K. Cerebellar mutism. Review of literature. Childs Nerv Sys 2011; 27:355-363.
7. Catsman-Berrevoets CE, Van Dongen HR, Mulder PG, Paz y Geuze D, Paquier PF, Lequin MH. Tumour type and size are high risk factors for the syndrome of "cerebellar" mutism and subsequent dysarthria. J Neurol Neurosurg Psychiatry 1999; 67:755-757.
8. Doxey D, Bruce D, Sklar F, Swift D, Shapiro K. Posterior fossa syndrome: Identifiable risk factors and irreversible complications. Pediatr Neurosurg 1999; 31:131-136.
9. Gelabert-González M, Fernández-Villa J. Mutism after posterior fossa surgery. Review of the literature. Clin Neurol Neurosurg 2001; 103:111-114.
10. Robertson PL, Muraszko KM, Holmes EJ, Sposto R, Packer RJ, Gajjar A, Dias MS, Allen JC; Children's Oncology Group. Incidence and severity of postoperative cerebellar mutism syndrome in children withmedulloblastoma: A prospective study by the Children's Oncology Group. J Neurosurg 2006; 105:444-451.
11. Van Calenbergh F, Van de Laar A, Plets C, Goffin J, Casaer P. Transient cerebellar mutism after posterior fossa surgery in children. Neurosurgery 1995; 37:894-898.
12. Wells EM, Walsh KS, Khademian ZP, Keating RF, Packer RJ. The cerebellar mutism syndrome and its relation to cerebellar cognitive function and the cerebellar cognitive affective disorder. Dev Disabil Res Rev 2008; 14:221-228.
13. Huber JF, Bradley K, Spiegler BJ. Dennis M. Long-term effects of transient cerebellar mutism after cerebellar astrocytoma or medulloblastoma tumor resection in childhood. Childs Nerv Syst 2006; 22:132-138.
14. Erşahin Y, Mutluer S, Cağlı S, Duman Y. Cerebellar mutism: Report of seven cases and review of the literature. Neurosurgery 1996; 38:60-65.
15. Aarsen FK, Van Dongen HR, Paquier PF, Van Mourik M, Catsman-Berrevoets CE. Long-term sequelae in children after cerebellar astrocytoma surgery. Neurology 2004; 62:1311-1316.
16. Erşahin Y, Yararbaş U, Duman Y, Mutluer S. Single photon emission tomography following posterior fossa surgery in patients with and without mutism. Childs Nerv Syst 2002; 18:318-325.
17. Huber JF, Bradley K, Spiegler B, Dennis M. Long-term neuromotor speech deficits in survivors of childhood posterior fossa tumors: Effects of tumor type, radiation, age at diagnosis, and survival years. J Child Neurol 2007; 22:848-854.
18. Steinbok P, Cochrane DD, Perrin R, Price A. Mutism after posterior fossa tumour resection in children: Incomplete recovery on long-term follow-up. Pediatr Neurosurg 2003; 39:179-183.
19. Ammirati M, Mirzai S, Samii M. Transient mutism following removal of a cerebellar tumor. A case report and review of the literature. Childs Nerv Syst 1989; 5:12-14.
20. Catsman-Berrevoets CE, van Breemen M, van Veelen ML, Appel IM, Lequin MH. Supratentorial arterial ischemic stroke following cerebellar tumor resection in two children. Pediatr Neurosurg 2005; 41:206-211.
21. Janssen G, Messing-Jünger AM, Engelbrecht V, Göbel U, Bock WJ, Lenard HG. Cerebellar mutism syndrome. Klin Padiatr 1998; 210:243-247.
22. Kotil K, Eras M, Akçetin M, Bilge T. Cerebellar mutism following posterior fossa tumor resection in children. Turk Neurosurg 2008; 18:89-94.
23. Morris EB, Phillips NS, Laningham FH, Patay Z, Gajjar A, Wallace D, Boop F, Sanford R, Ness KK, Ogg RJ. Proximal dentatothalamocortical tract involvement in posterior fossa syndrome. Brain 2009; 132:3087-3095.
24. Puget S, Boddaert N, Viguier D, Kieffer V, Bulteau C, Garnett M, Callu D, Sainte-Rose C, Kalifa C, Dellatolas G, Grill J. Injuries to inferior vermis and dentate nuclei predict poor neurological and neuropsychological outcome in children with malignant posterior fossa tumors. Cancer 2009; 115:1338-1347.
25. Riva D, Giorgi C. The cerebellum contributes to higher functions during development: evidence from a series of children surgically treated for posterior fossa tumours. Brain 2000; 123:1051-1061.
26. Kusano Y, Tanaka Y, Takasuna H, Wada N, Tada T, Kakizawa Y, Hongo K. Transient cerebellar mutism caused by bilateral damage to the dentate nuclei after the second posterior fossa surgery. Case report. J Neurosurg 2006; 104:329-331.
27. McMillan HJ, Keene DL, Matzinger MA, Vassilyadi M, Nzau M, Ventureyra EC. Brainstem compression: A predictor of postoperative cerebellar mutism. Childs Nerv Syst 2009; 25:677-681.
28. Caner H, Altınörs N, Benli S, Calışaneller T, Albayrak A. Akinetic mutism after fourth ventricle choroid plexus papilloma: treatment with a dopamine agonist. Surg Neurol 1999; 51:181-184.
29. D'Avanzo R, Scuotto A, Natale M, Scotto P, Cioffi FA. Transient "cerebellar" mutism in lesions of the mesencephalic-cerebellar region. Acta Neurol (Napoli) 1993; 15:289-296.
30. Siffert J, Poussaint TY, Goumnerova LC, Scott RM, LaValley B,Tarbell NJ, Pomeroy SL. Neurological dysfunction associated with postoperative cerebellar mutism. J Neurooncol 2000; 48:75-81
31. Aguiar PH, Plese JP, Ciquini O, Marino R. Transient mutism following a posterior fossa approach to cerebellar tumors in children: a critical review of the literature. Childs Nerv Syst 1995; 11:306-310.
32. Özgür BM, Berberian J, Aryan HE, Meltzer HS, Levy ML. The pathophysiologic mechanism of cerebellar mutism. Surg Neurol 2006; 66:1-25.
33. Dietze DD Jr, Mickle JP. Cerebellar mutism after posterior fossa surgery. Pediatr Neurosurg 1990; 16:25-31.
34. El-Bahy K. Telovelar approach to the fourth ventricle: Operative findings and results in 16 cases. Acta Neurochir (Wien) 2005; 147:137-142.
35. Grill J, Viguier D, Kieffer V, Bulteau C, Sainte-Rose C, Hartmann O, Kalifa C, Dellatolas G. Critical risk factors for intellectual impairment in children with posterior fossa tumors: the role of cerebellar damage. J Neurosurg 2004; 101:152-158.
36. Mussi AC, Rhoton AL Jr. Telovelar approach to the fourth ventricle: Microsurgical anatomy. J Neurosurg 2000; 92:812-823.
37. Rajesh BJ, Rao BR, Menon G, Abraham M, Easwer HV, Nair S. Telovelar approach: technical issues for large fourth ventricle tumors. Childs Nerv Syst 2007; 23:555-558.
38. Tanrıöver N, Ulm AJ, Rhoton AL Jr, Yasuda A. Comparison of the transvermian and telovelar approaches to the fourth ventricle. J Neurosurg 2004; 101:484-498.
39. von Hoff K, Kieffer V, Habrand JL, Kalifa C, Dellatolas G, Grill J. Impairment of intellectual functions after surgery and posterior fossa irradiation in children with ependymoma is related to age and neurologic complications. BMC Cancer 2008: s. 8:15.