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Ramsay Hunt syndrome revisited-emphasis on Ramsay Hunt syndrome with multiple cranial nerve involvement

Eva Rye Rasmussen[1*], Eva Lykke[2], Jan Gren Toft[3] and Kristianna Mey[4]

*Correspondence: eva.rye.rasmussen@dadlnet.dk

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[1]Department of Otolaryngology-head and neck surgery, Lykkebaekvej 1, 4600 Koege, Denmark.
[2]Department of Otolaryngology-head and neck surgery, Blegdamsvej 9B, 2100 Copenhagen East, Denmark.
[3]Private practice Ear, nose and throat diseases Strandgade 41, 4800 Nykoebing Falster, Denmark.
[4]Department of Otorhinolaryngology and audiology, Copenhagen University Hospital Rigshospitalet/Gentofte, Niels Andersens vej 65, 2900 Hellerup, Denmark.

Abstract

Introduction: The Ramsay Hunt syndrome is characterized by herpetic lesions combined with peripheral facial nerve palsy. The disease is caused by a reactivation of the varicella zoster virus and can be deceiving since the herpetic lesions are not always present (zoster sine herpete) and might mimic other severe neurological illnesses. This article reviews the various forms of Ramsay Hunt syndrome and how they can give rise to diagnostic and therapeutic challenges.

Material and method: Studies on the assessment and treatment of Ramsay Hunt syndrome were found by conducting a thorough literature search in PubMed, Medline, The Cochrane Library database and Google Scholar using the search words «varicella», «zoster», «ramsay hunt», «oticus», «cranial nerve», «facial nerve» and combinations thereof. The bibliographies of substantial articles were subsequently assessed.

Results: About 12% of all peripheral facial nerve palsies are caused by varicella zoster virus. In more than 50% pain is the initial symptom making the diagnosis difficult. Female gender, and in general age above 50 years, renders patients more susceptible to Ramsay Hunt syndrome. The main prognostic factor is the severity of the initial symptoms. The occurrence rate of associated cranial polyneuropathy has been reported to be 1.8-3.2% and cranial nerves VII, VIII, IX are the ones most commonly affected. The full recovery rate is reported to be as low as 27.3% when multiple cranial nerves are involved. Combination therapy comprising of antiviral drugs and corticosteroids is recommened and should be initiated within 72 hours. Vaccination against varicella zoster virus is an interesting new development that might reduce the incidence of varicella zoster virus associated disease altogether.

Conclusion: Ramsay Hunt syndrome is a difficult and severe diagnosis with a low full recovery rate. Extensive randomized trials are urgently needed to verify the optimal treatment and the efficacy of varicella zoster virus vaccine in both naïve and herpes zoster patients.

Keywords: Ramsay Hunt syndrome, facial paralysis, herpes zoster, herpes zoster oticus, cranial nerve diseases

Introduction

James Ramsay Hunt was an American physician and scientist who, in the 20th century, described a series of cases suffering from herpetic lesions in the oral cavity or external ear, combined with facial palsy and often accompanied by other neurological disturbances [1]. Since those days, the Ramsay Hunt syndrome (RHS) has been investigated thoroughly and different classifications and definitions of the disease have been proposed [1,2]; however no single definition has been agreed upon representing a 'golden standard'. Hunt himself classified the disease into four subgroups according to the extent of the pathological processes taking place in the geniculate ganglion (**Table 1**) [3]. The disease is caused by a reactivation of the varicella zoster virus (VZV) of the alpha herpes family; the symptoms are deceptive since the herpetic lesions are not always present (zoster sine herpete) and might mimic several other neurologic illnesses such as cerebral insults, Lyme disease and meningitis. This article reviews the various forms of RHS in adult immunocompetent patients as they present themselves to a broad selection of medical specialties, leading to diagnostic and therapeutic challenges.

Method

In order to evaluate earlier work with regard to the assessment and treatment of RHS, a thorough literature search was conducted in PubMed, Medline, The Cochrane Library database and Google Scholar using the search words «varicella», «zoster», «ramsay hunt», «oticus», «cranial nerve», «facial nerve» and combinations thereof. A PubMed search exclusively on «Ramsay Hunt» alone produced 431 hits; the search for «zoster oticus» produced 407 hits, thus proving the need for a more restrictive search strategy. Only articles in English published

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Table 1. Original classification of Ramsay Hunt syndrome.

Hunt's classification of 'his' syndrome
1. Disease affecting the sensory portion of the seventh cranial nerve
2. Disease involving the sensory and motor divisions of the seventh cranial nerve
3. Disease affecting the sensory and motor divisions of the seventh cranial nerve with auditory symptoms
4. Disease affecting the sensory and motor divisions of the seventh cranial nerve with both auditory and vestibular symptoms

since the year 2003 were included in the subsequent search; resulting in 82 hits. In order to find the original articles, the bibliographies of selected reviews were examined, expanding both the time frame and the allowed languages (English, German, Norwegian and Danish). Articles reporting solely on immune-incompetent patients were excluded. Patients suffering from diabetes mellitus were regarded as immune-competent in this review. In some articles both immune-competent and–incompetent patients were reported on, but for our purposes the immune-incompetent patients has been excluded. Furthermore the authors prioritized original research rather than reviews and reference articles. The search was concluded on 11th of April 2013.

Epidemiology

General data and incidence

About 12% of all peripheral facial nerve palsies are caused by VZV [4]. Female gender, and in general age above 50 years, renders patients more susceptible to Ramsay Hunt syndrome [5,6]. Epidemiological data is summarized in **Table 2** [6-8]. The reason for female predisposition to Ramsay Hunt syndrome is unknown, but the incidence of i.e., herpes simplex infection is also higher in females [9]. Regarding the frequency of RHS and cranial nerve involvement the statements in the literature diverge substantially. This might be due to the fact that some neurological deficits are difficult to assess, especially for the untrained eye. Since RHS is a relatively rare condition, not much epidemiological data is available on which cranial nerves are most commonly affected (**Table 2**). The occurrence rate of associated cranial polyneuropathy has been reported to be 1.8-3.2% [5,10,11]. We gathered information on the involvement of cranial nerves from case reports published from 2003 to 2013 on immunocompetent adults (**Table 3**) [12-36].

Prognosis

The rate of full recovery was found to be 67.7% in RHS, but as low as 27.3% in patients with multiple cranial nerve involvement [10]. By contrast the data on recovery rate for individual cranial nerve involvements are not well established [10,37]. Post-herpetic neuralgia has been reported in as many as 50% of RHS patients [38]. The main prognostic factor seems to be the severity of the initial symptoms [39]. The US and Japan have now implemented varicella zoster vaccination as part of their childrens vaccination programme and the incidence of chickenpox, herpes zoster (HZ) and sequelae has declined significantly [6,40,41]. In persons 60 years of age or older, a reduction in incidence of HZ and post-herpetic pain of more than 50% has been shown to be achievable by vaccination [42].

Table 2. Epidemiological data and prognosis [5-7,10,33,38,40,42].

Epidemiological Data	
Incidence of HZ:	HZ oticus: 1% of which 96% are associated with peripheral facial nerve palsy
	HZ in general: 3.6 per 1000 person-years, 3% with neurological complications
Incidence of involved cranial nerves:	HZ in general: Ophthalmic nerve affected in 10-29% and facial nerve affected in 1%
	Associated cranial polyneuropathy : 1.8-3.2%
	VII, VIII, IX, X, V, III/XII (incidence in decending order)
	VII, VIII, IX, V, X, VI, III, XI, IV, I/II/XII (incidence in decending order)
Prognosis/recovery:	Full recovery: 45.5% of facial palsy and 11.1% of hearing loss
	Full recovery: 46.7% of auditive symptoms
	Post-herpetic neuralgia in 50% of RHS patients
Introduction of Varicella Zoster vaccine	Incidence of chickenpox, HZ and sequelae has declined
	Incidence of HZ and post-herpetic pain reduced with more than 50%
	Decreased incidence of HZ (and thus RHS) in vaccinated populations

Pathophysiology

Natural history of VZV

VZV, as does the herpes simplex virus, belongs to the family of alpha herpes viruses. It infects almost all humans in the form of chickenpox (varicella) and is spread by aerosolized respiratory droplets from infected individuals or by direct contact with a herpes zoster rash. The incubation period is 10-21 days and patients are contagious from 2 days before the rash appears until all elements have crusted, usually after 5-7 days. HZ is likewise contagious. VZV is a neurotrophic virus causing the classical variety of chickenpox in person's primary infected. Children usually exhibit milder symptoms than do adults, mainly manifested as an itching vesicular cutaneous rash; however severe and even fatal complications have been reported. Using the host's immune system, especially its T-cells, the virus is delivered to the skin and sensory nerves [43,44]. The virus is never cleared from the body but establishes a latent infection in the sensory ganglia and the facts that it does not spread is due to host's immune response and VZV antibodies. In the ganglia, the virus keeps itself hidden from the host's immune system [45,46]. Later in life the virus, like other herpes

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Table 3. A review of reported cases with immunocompetent adults diagnosed with Ramsay Hunt syndrome combined with multiple cranial nerve involvement from 2003-2013. Cranial nerve involvement in descending order of occurrence in parenthesis: VII(27), VIII(26), X(10), V+ IX(8), XII(7), XI+IV(2) and I+III+VI(1) [12-36].

Author & publication year	Number of patients in study	Affected cranial nerves	Age In years	Sex Male/Female
Kim D / 2008	1	VII+VIII	51	F
Liao / 2011	1	VII+VIII	60	M
Bhagra / 2006	1	VII+VIII	64	F
Gondivkar / 2010	1	VII+VIII	32	M
Galimi / 2011	1	VII+VIII	48	M
Van de Steene / 2004	2	VII+VIII VII+VIII	64 58	F M
Syal / 2004	1	VII+VIII (bilateral)	57	M
Hung / 2010	1	V+VII+VIII	77	M
Akyol / 2006	1	V+VII+VIII	61	M
Lim / 2011	1	V+VII+VIII	50	F
Padhiary / 2007	1	VII+VIII+IX+X	44	M
Wang / 2011	1	VII+IX+X	25	M
Saito / 2003	2	VII+VIII VII+VIII+X	39 66	M F
Kim JH / 2007	1	VII+VIII+IX+X	27	M
Shim / 2011	1	VII+VIII+IX+X	48	M
Espay / 2005	1	VII+VIII+IX+XII	83	F
Izumi / 2007	1	VII+VIII+XII	72	M
Kim TU / 2012	1	VII+VIII+IX+X+XII	73	F
Lauridsen / 2010	1	V+VII+VIII+X+XII	56	M
Sun / 2011	1	V+VII+VIII+XII	62	F
Coleman / 2012	1	V + VII + VIII + X	81	F
Godani / 2013	1	IV + VII+ VIII	69	F
Nishioka / 2006	1	III+IV+V+VI+VII+VIII +IX+X+XI+XII	78	M
Morelli / 2008	1	V+VII+VIII+IX+X+XI+XII	82	M
Sims / 2008	1	I+VII+VIII	81	M

viruses, can become re-activated causing zoster (shingles) in all parts of the body [43]. The reason for the re-activation is believed to be caused by lessened immunity in the host over time [42,43,47]. For the varicella zoster life-cycle see **Figure 1**.

Localization of virus during zoster

Different theories have been proposed as to where exactly VZV and associated inflammation are situated during a flare up of herpes zoster, but generally it is acknowledged, that the cause is ganglionitis. Theories involving viral spread due to vasculitis or perineuritis have also been considered [34,48] and VZV has been electron microscopically observed in sensory nerves during HZ [49]. VZV is also found in the zoster elements during the vesicular phase [44]. Usually, no radiologic assessment is necessary, but Magnetic resonance imaging studies using gadolinium-diethylenetriamine penta-acetic acid often show enhancement of the intra-temporal segments of the facial nerve and internal auditive canal in patients with RHS [50].

Clinical presentation

RHS is a diverse and challenging disease, since it can be associated with neurological symptoms mimicking other diseases. Pathognomonic for the syndrome are peripheral facial nerve palsy and vesicular rash on the external ear, external auditory canal (which might only be visible by otoscopic assessment), and/or the mucosa of the ipsilateral part of the tongue or palate, usually accompanied by otalgia (**Figures 2a** and **2b**) [1,7]. By contrast there are also reports on facial nerve palsy and other cranial nerve palsies without herpetic lesions but with an increase of VZV antibodies, so called zoster sine herpete [51-53]. Pain in the affected area develops prior to a vesicular rash in 14-50% of VZV patients; the pain may mimic

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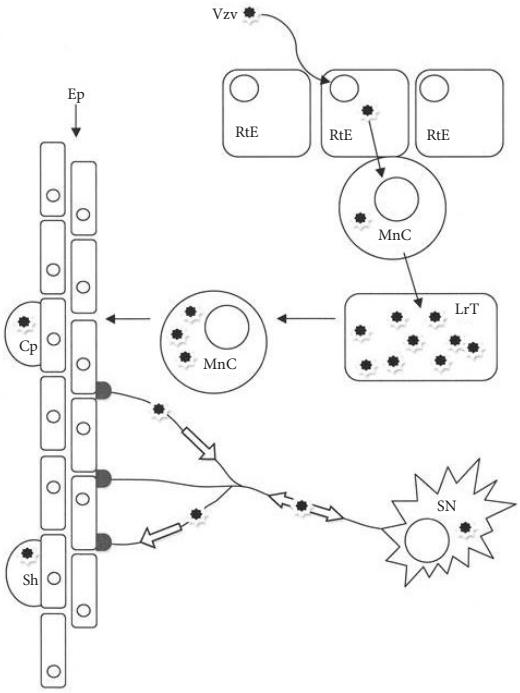

Figure 1. The Varicella Zoster Virus life cycle.
The varicella zoster virus (Vzv) enters the body through the respiratory tract epithelium (RtE). From here it infects the mononuclear cells (MnC), especially T-cells that carry the virus to the lymphoreticular tissues (LrT) where it replicates. In association with the MnC the virus enters the blood stream causing viremia and is seeded out to epithelial sites (Ep) creating chickenpox (Cp). The mucocutaneous lesions heal but the virus enters sensory nerve endings, is transported via the peripheral nerve to the sensory ganglia and a latent infection is established. Later in life a secondary infection can occur causing zoster or shingles (Sh) in the dermatome at the site of reactivation.

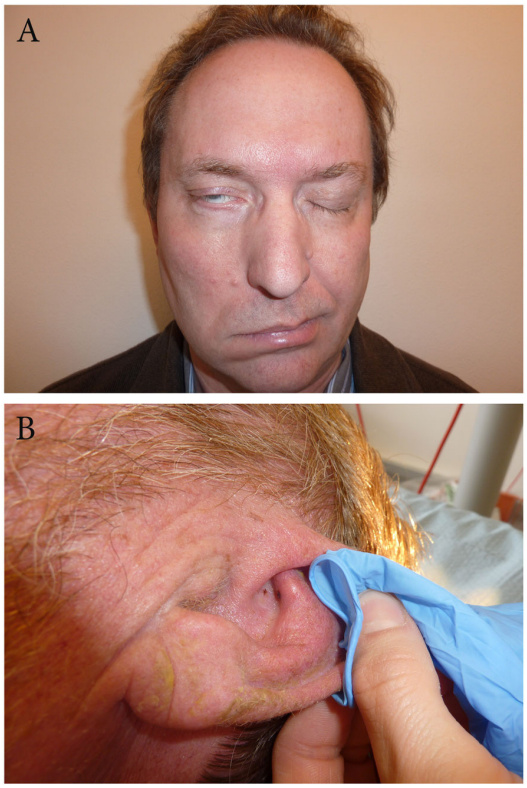

Figure 2. Peripheral facial nerve palsy and vesicular rash on the auricle. (**A**) Peripheral facial nerve palsy in a patient suffering from Ramsay Hunt syndrome. (**B**) Vesicular rash of the auricle in the remission phase.

acute otitis and other ear diseases [6,39]. Furthermore, facial nerve palsy might develop several days before the rash in up to 20% of RHS patients [11,39]. More than half the patients have pain as their first symptom while as few as 2% have a rash as the initial symptom [39]. The usual RHS patient and the possible associated symptoms are described in **Table 4**.

Diagnostic approach

The RHS diagnosis is purely clinical, but in some cases a blood test for VZV antibodies may be useful. When other cranial nerves are affected, Magnetic resonance imaging may be necessary to exclude intracerebral pathology. The House Brachmann scale should be used to assess the facial nerve function initially and at every subsequent follow-up.

Treatment

Antiviral drugs and/or corticosteroids

RHS being a rare condition, only few studies on how to treat RHS have been carried out. Most of these studies are in favor of

Table 4. Pathognomonic and secondary symptoms of the Ramsay Hunt syndrome and related cranial nerves. Incidence in descending order.

Symptoms	Affected cranial nerves
Pathognomonic symptoms (always present)	
Herpetic vesicular rash	VII
Peripheral facial nerve palsy	VII
Secondary symptoms (might be present)	
Otalgia	V
Loss of hearing	VIII
Hyperacusis (due to palsy of chorda tympani)	VII
Vestibular symptoms (dizziness, nystagmus)	VIII
Taste disturbances, reduction of tear secretion	VII
Hoarseness (vocal cord palsy)	X
Weakness of the masseter and temporal muscles	V
Asymmetrical rise of the soft palate and/or uvula deviation	IX
Dyssensitivity of the face	V
Tongue deviation	XII
Dysphagia	IX, X
Ocular deviation	III,IV,VI
Dilated pupil	III

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the use of antiviral drugs [54,55]. According to a Cochrane review, only one randomized controlled trial was performed including 15 patients; and no significant conclusions could be drawn as the study was deemed of low quality [56]. Other studies comprising small study populations have shown increased effects of acyclovir combined with corticosteroids compared to corticosteroids alone [39,57]. A main prognostic factor on outcome, with regard to medication, seems to be the time from debut of symptoms to initiation of treatment, whether consisting of combined acyclovir and corticosteroids or corticosteroids alone (Table 5) [58]. Some studies report on other treatment regimens like bed-rest, high protein, low sodium diet, peripheral vasodilators combined with acyclovir and corticosteroids, surgical decompression or anastomosis of the facial nerve but more extensive and randomized controlled studies are needed to define a 'golden standard' of RHS treatment [10,59]. Until those studies are available, acyclovir in combination with corticosteroids are considered the correct treatment of RHS.

Vaccination

For persons of 60 years or older, vaccinating them against VZV effectively reduces the risk of HZ and post-herpetic pain whether or not these persons had had prior exposure to VZV. The pathophysiology is ascribed to the booster effect of the cell-mediated immunity against the dormant VZV (42). In children, the course of primary varicella zoster infection (chickenpox) in unvaccinated patients is markedly milder when vaccinated within 3 days from exposure [60,61]. Vaccinations are well tolerated and result in a 2.1 fold increase in VZV antibody level in patients with a positive history of HZ [62]. To the best of our knowledge, no substantial studies on the prognosis of the clinical outcome regarding vaccination or passive immunization of immunocompetent HZ or RHS patients have

Table 5. Recent recommendations in the treatment of Ramsay Hunt syndrome.

Study/year	No. of Patients	Combination therapy	Antiviral treatment	Corticosteroid treatment	VZV Vaccine	Initiation of treatment (d=days)	Comments / strong points of study
Coulson 2011	101	YES	From 1998, famciclovir 250mg x 3, 21d	1mg/kg/d, 14 d, decreased by 10mg/d until 0	NC	Early:< 5 d Late: > 5 d	Statistically significantly better in combination treatment of early ACY+late COR
Uri 2003	31	YES	ACY 15mg/kg/d for 7 d	100mg x 3 for 7 d	NC	Early 1-3 d Late 4-7 d	Recommends IV ACY over oral administration
Kinishi 2001	91	YES	ACY 4000 mg/d for 7d	500→100 mg/d decreased over 7d	NC	< 7 d	Assess ACY+COR on facial nerve recovery compared to COR alone
Hato 2000	52	NC	NC	NC	YES	NC	Reviews prognostic factors for children with RHS. None of the 52 had VZV vaccine; the incidence of RHS in later years drops
Murakami 1997	80	YES	ACY either 250mg x 3 IV or 800mg x 5 orally for 7 d	1mg/kg/d for 5 d, decreased 10 mg/d until 0, over 10 d	NC	Early 1-3 d Late >7 d	75% recovered in 1-3 d group 30% recovered in >7 d group Argues that onset of treatment is essential
De RU 2011	NA	YES	NA	NA	NC	NC	Meta-analysis of reviews 1985-2010 – strongly recommend combination therapy
Uscategui 2008 (Oct)	NA	No evidence	NA	NA	Maybe	< 3 d	Cochrane Review; finds no quality RCT to support the use of antiviral compared to the use of COR alone
Uscategui 2008 (June)	NA	No evidence	NA	NA	Maybe	< 3 d	Cochrane Review; finds no RCT at all to address use of COR to support antivirals

ACY: Acyclovir, COR: Corticosteroids, VZV: Varicella zoster virus, NC: Not commented, NA: Not applicable, RCT: randomized controlled trials, IV: Intravenously, d: days

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been carried out. In theory, while vaccination of RHS patients at initial presentation could have some effect, this effect should be compared to the efficacy of corticosteroids and acyclovir.

Conclusion

RHS is a rare but severe condition defined by herpetic rash of the ear or the mucosa of the mouth and peripheral facial nerve palsy, most often combined with otalgia. The syndrome can be associated with several other cranial nerve symptoms mimicking intracerebral disease. The full recovery rate is reported to be as low as 27.3%. Antiviral therapy combined with corticosteroids is recommended by most scientists in the field, although more randomized controlled trials are needed. Vaccination against VZV is an interesting new development that might reduce the incidence of VZV associated disease altogether. More studies regarding the potential efficacy of the VZV vaccine, given at the early sign of RHS, are urgently needed to combat this severe disease.

List of abbreviations

RHS: Ramsay Hunt syndrome
VZV: Varicella zoster virus
HZ: Herpes zoster

Competing interests

The authors declare that they have no competing interests.

Authors' contributions

Authors' contributions	ERR	EL	JGT	KM
Research concept and design	✓	--	--	✓
Collection and/or assembly of data	✓	✓	✓	✓
Data analysis and interpretation	✓	✓	--	✓
Writing the article	✓	✓	✓	✓
Critical revision of the article	✓	--	--	✓
Final approval of article	✓	✓	✓	✓

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