زراعة الحلزون في تشوه الجوف الواحد باستخدام الجهاز المفصل خصيصاً حسب حجم الجوف

جمال قسومة*

الملخص

هدف البحث: هو تقييم النتائج السمعية والكلامية لعمليات زرع الحلزون في تشوه الجوف الواحد إذْ إنَّ الإذن الداخلية توقفت عن التطور في الأسبوع الثالث من الحياة الجنينية. وكان في الماضي يعدُّ وجود أي تشوه في الحلزون هو مضاد استطباب للعملية، ولكن بدأت تجارب في بعض المراكز وحصل بعض النجاح مما شجع على متابعة العمل في بقية المراكز وشجعني على إجراء هذه العمليات الثلاث.

طريقة البحث: أجريت دراسة استباقية: قُيِّمَ المرضى بشكل دوري من قبلي ومن قبل مدربة النطق ومختص السمعيات ومختص البرمجة ومن خلال استجواب الأهل على مدى أداء الطفل في المنزل. ثم جُمِعَت المعلومات وصُنِّفَتْ حسب التصنيفات العالمية لتمييز الكلام سمعياً الذي هو ثماني درجات بدأت من "عدم الإلمام نهائيا بالأصوات المحيطة وانتهت ب"استعمال الهاتف مع شخص معروف للمريض" ثم أخذ الوسطي منها. وقُورنَ بنتائج مركز عالٍ معروف.

* قسم الأذنية- كلية الطب البشري- جامعة دمشق.

ثم درست إمكانية فهم كلام المريض من قبل الآخرين وهو ست درجات من "كلام غير مفهوم" إلى "كلام مفهوم لمعظم الناس" وأخذ الوسطي منها، وقُورنَ مع مركز عالمي معروف.

النتائج: أُخذَ الوسطي من حيث تمييز الكلام سمعياً وفهم الكلام وقُورنَ مع مركز مقاطعة يورك في بريطانيا فوجد أن تطورهم أقل من الأطفال الطبيعيين وخاصة في الأشهر الستة الأولى، ولكن بعد ذلك كانت النتائج متقاربة.

الاستنتاجات: إن وجود تشوه في الحلزون لم يعد يعدُّ مضاد استطباب للعمل الجراحي كما كان يعتقد وقد أظهرت النتائج ما يؤكد ذلك.

الكلمة المفتاح: زراعة الحلزون – تشوه الجوف الواحد.

Cochlear Implant in Common Cavity

Jamal Kassouma*

Abstract

Objective: to evaluate the results of cochlear implant operations performed on patients with common cavity anomalies in the inner ear; whereas, the ear stopped developing at the end of the third week. In the past, the presence of any anomaly in the cochlea was considered a counter-indication for surgery but experimentation has been carried out worldwide and there were some major breakthroughs, which encouraged other centers worldwide to carry out this surgery and I was encouraged myself to perform 3 surgeries, which is a fair number, considering that the number of operations performed worldwide, as to date, is around 20.

Means of Research: Prospective: The patients were routinely evaluated by the audiologists, speech pathologists and myself and also through interviewing the patient's family about the child's performance at home. The data was then gathered, analyzed and categorized according to international standardization of auditory recognition, which is divided into separate categories, which range from "inability to recognize environmental sounds" to "effective use of the telephone with a known person".

*Lecturer and Consultant ENT Surgeon Faculty of Medicine, Damascus University.

These results were then averaged and compared to the Yorkshire Cochlear Implant Center, U.K. Then the patients' speech intelligibility after CI operations was evaluated. The results were then gathered and organized according to the international standardization of speech intelligibility which ranges from "unintelligible speech" to "speech that can be understood by most people". These results were then averaged and compared to those of the YCIC.

Results: The results were averaged and compared to these of the YCIC for auditory recognition and speech intelligibility and it appears that these children's results were below those of children without these anomalies, especially during the first six months post-op., but afterwards, the results were close.

Conclusion: The presence of anomalies in the cochlea is no longer considered a counter-indication for surgery as was previously known, as the above results confirm.

Key Word: Cochlear implantation – common cavity deformity .

Patients with congenital malformation of the inner ear are being considered for cochlear implantation in increasing numbers surgery was not difficult as the cavity was located in the location of the lateral semicircular canal in normal ear. we report the results of three cases and measured the auditory performance and speech recognition over time, then we compared it with the results of YCIS (Yorkshire Cochlear Implant Service), UK, for 65 children with normal cochlea. We concluded that the results were remarkably similar after 18 months.

The transmastoid facial recess approach has become the standard technique for cochlear implantation. Although this approach has been used for implantation in patients with common cavity deformities, it is not without increased risk to the facial nerve. Using a direct approach to the common cavity we have successfully implanted three patients with common cavity deformities using Med El custom- made devices.

Jackler et al(1) suggested a new classification of the inner ear malformations based logically, on the stage of embryonic life at which development appears to have been arrested.

1. Complete aplasia (Mochel)
2. Cochlear aplasia – semi-circular canals and vestibular normal or malformed.
3. Cochlear hypoplasia: small cochlear bud.
4. Common cavity: single chamber cochlea
5. Incomplete partition of cochlea (Mondini)

Although radiologically apparent Mondini's deformities account for the majority of the cochlear inner ear anomalies, the second most frequently encountered anomaly in a study by Jackler and Luxford was the common cavity malformation; the common cavity anomaly is thought to result from an arrest in differentiation of the otocyst during the fourth gestational week, whereas the Mondini deformity results from an arrest is development during the seventh gestational week, as a consequence, the degree of hearing loss associated with common cavity is usually greater than the loss associated with Mondini's malformation, making individuals with common cavity deformities more likely candidates for cochlear implantation, despite the severity of cochlear malformation in this deformity, a sufficient number of ganglion cells exists making these patients suitable for cochlear implantation. Linthicum et al. (2) reported

on the temporal bone finding of a patient able to perceive auditory percepts with as few as 3300 spiral ganglion cells.

Reports by Migamota et al. (3) Jackler et al. and Molter et al. (4) confirm the ability of the patients with common cavity deformities to benefit from cochlear implantation.

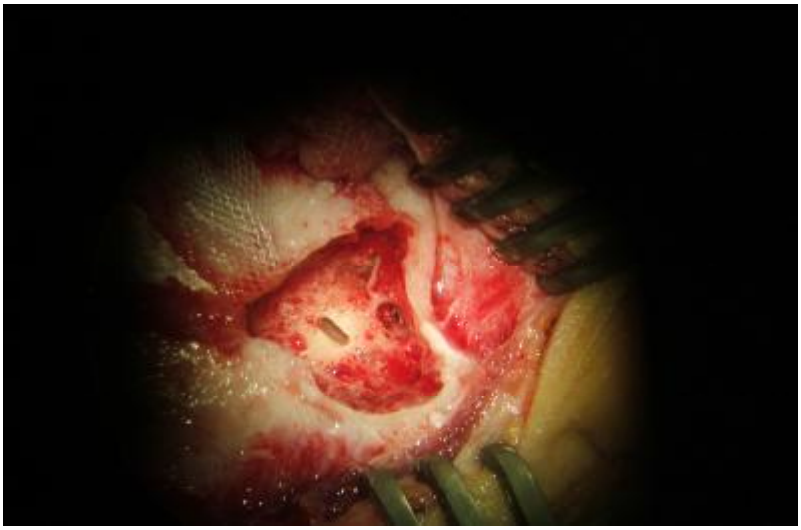
We implanted three cases with common cavity, the follow up was between six and eighteen months, and the progress was similar to that of the normal cochlea implanted children.

Surgical Technique:

Post aural incision posterior superiorly was carried out. A standard cortical mastoidectomy is performed, being careful not to saucerize the margins of the mastoidectomy. The sigmoid sinus is skeletonized; as in the middle fossa plate. Using a combination of cutting and diamond burs, the pneumatized bone is removed to facilitate visualization of the aditus and antrum. The pneumatized bone is removed posterior and inferior to the aditus, revealing the endochondral bone of the common cavity. The facial recess is opened in two cases to visualize: the stapedus muscle and to do stapedial reflexes.

Once the labyrinthine bone is clearly delineated in the area of the lateral semicircular canal; a slit labyrinthotomy is created using diamond buns. Once the endosteum of the common cavity is opened, the cavity was dry in our three cases, while McElveen (5) reported a free flow of spiral fluid in his cases. A Med-El custom made device was implanted. Telemetry was ok in all channels for three cases; we managed to see some stapedial reflexes in 2 cases where facial recess was exposed.

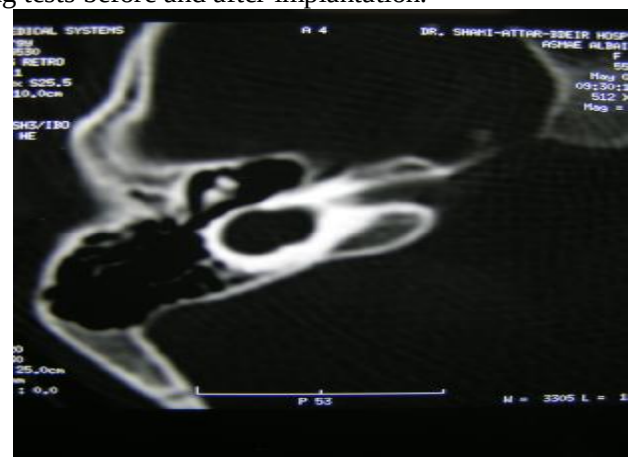
Labyrinthotomy was sealed with facia.. Intra-operative facial nerve monitoring is used in all cases.

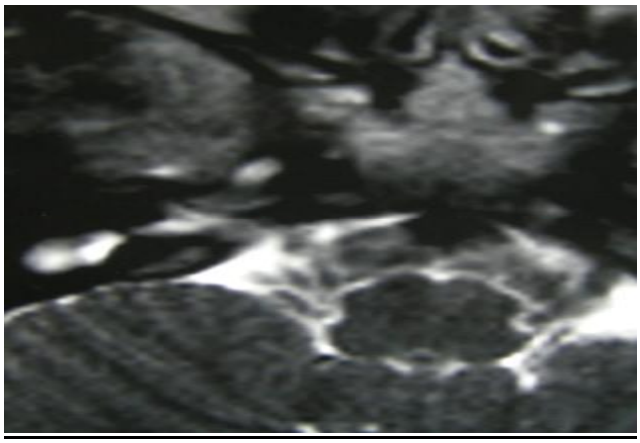


Case 1:

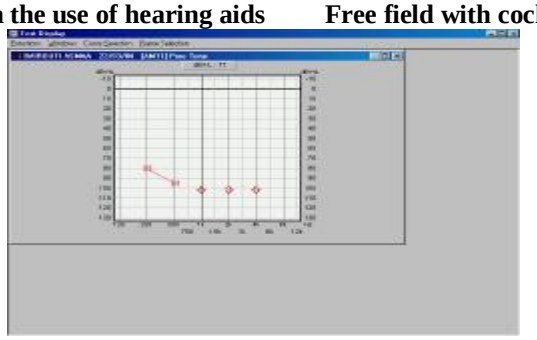
A.B. is a 2 year, 7 month old girl who noted to be deaf at the age of 7 months subsequent audiological testing revealed profound sensorineural hearing loss; she has a deformity of the eyelids. CT scan showed bilateral

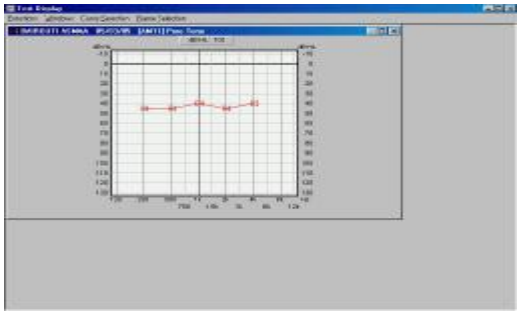
common cavity deformities. MRI showed 2 nerved in the IAC. She was implanted with Med-El custom made device, posterior tympanotomy was carried out and we managed to get stapedial reflexes for the electrode 2-6. The cavity was dry and the facia was packed around the electrode array at the insertion site. The surgery was without complication and she was discharged on the next day, a month later 12 electrodes were stimulated resulting in auditory perception and she developed some words very soon after implantation. We will demonstrate her CT, MRI, post implant XR, her hearing tests before and after implantation.





Free field with the use of hearing aids **Free field with cochlear implant**

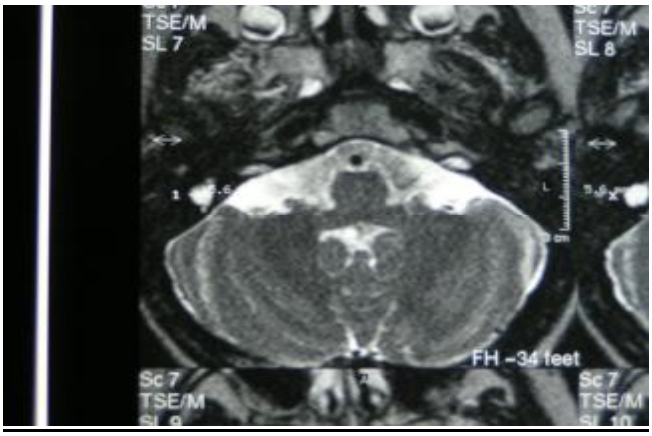


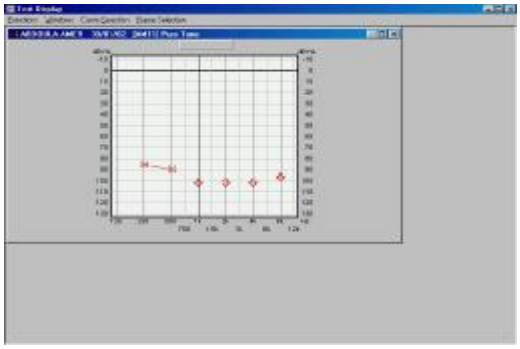


Case 2:

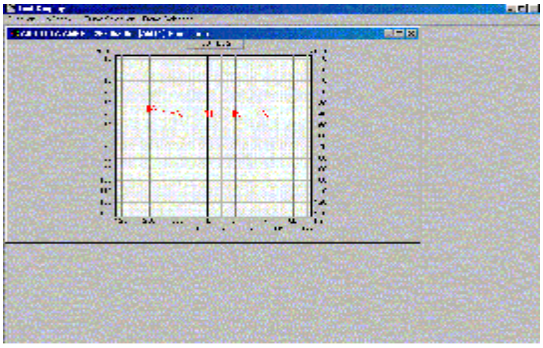
A.A. is a 6 year old boy with a profound sensorineural hearing loss. CT scan showed bil common cavity deformities, MRI showed 2 nerves in the

IAC and nerve fibres attached to the cavity; she was implanted with Med-El custom made device. Stapedial reflexes were positive for electrode 4.5.6. The cavity was dry. The surgery was without complications, a month later 12 electrodes were stimulated resulting in auditory perception and he developed some word very soon after implantation and a year later could speak a sentence. We will demonstrate his CT, MRI, post implant XR, his hearing tests before and after implantation.





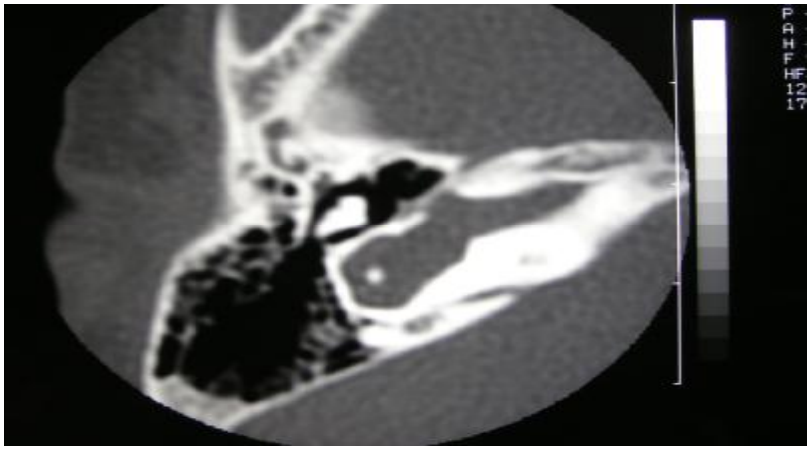
Free field with use of hearing aids

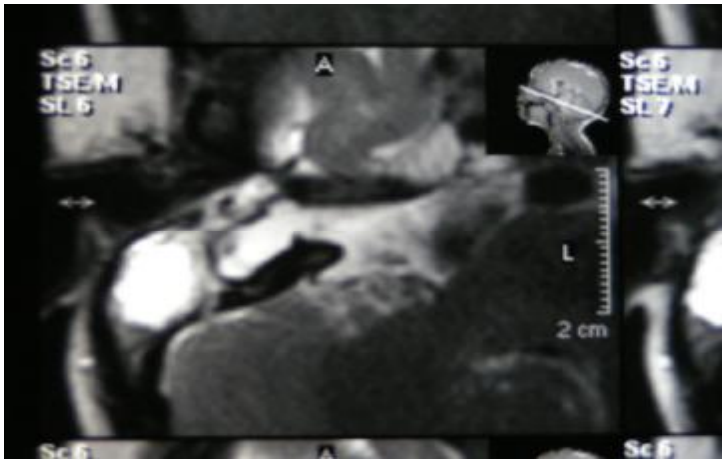


Free field with cochlear implant

Case 3:

Y.A. is a 2 year old girl with a profound sensorineural hearing loss. CT scan demonstrated bilateral common cavity deformities. MRI showed some nerve fibers attached to the cavity and 2 nerves in the IAC. She was implanted with Med-El custom made device. The cavity was dry and the surgery was without complications. A month later 12 electrodes were stimulated. We will demonstrate her CT, MRI, post implant XR her hearing tests before and after implantation.

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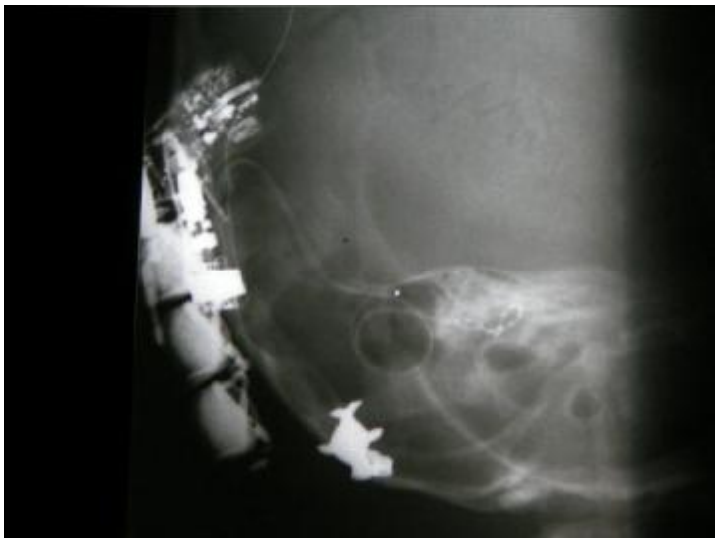
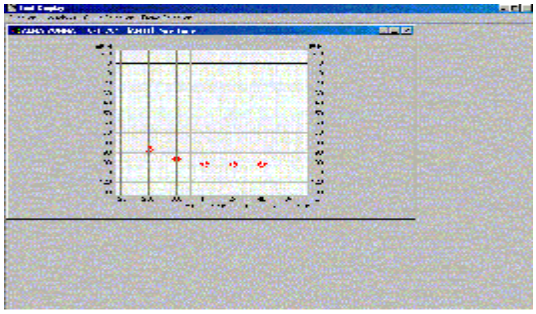
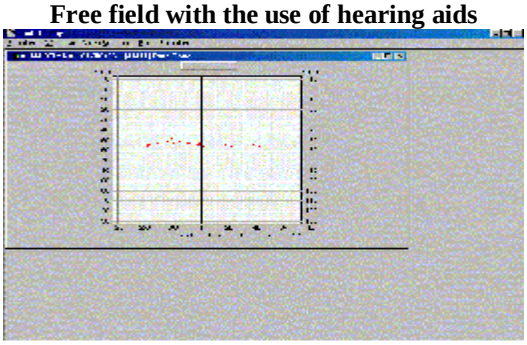




Free field with the use of hearing aids



Free field with cochlear implant

Discussion:

Transmastoid labyrinthotomy approach used in four patients by McElveen for electrode array placement and it was first performed in 1990 by one of the authors JMT and reported by Molter et al. few surgeon have used this approach.

Common Cavity Literature Review

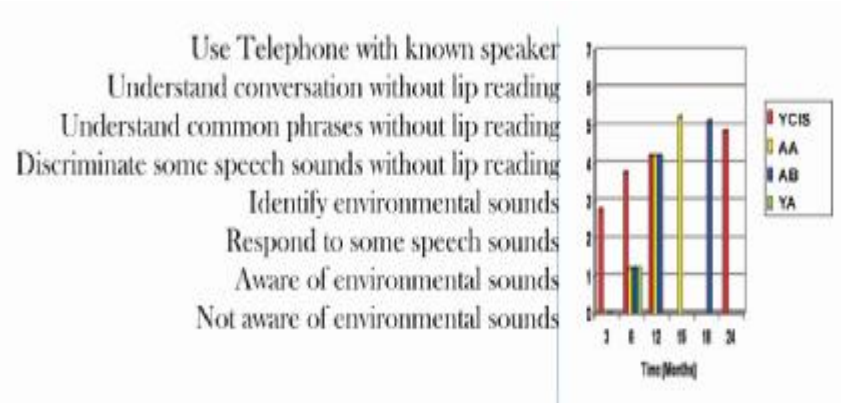
Authors	Patients(n)	Age	Device	Cochlear Configuration	CSF Leak	Operative Findings
Miyamoto et al.	1	Child	Nucleus	Common cavity	+	None (facial recess)
Jackler and Luxford	1	Child	3M/House	Common cavity	-	None (facial recess)
Motler et al.	1	Child	Nucleus	Common cavity	-	Aberrant VII labyrinthotomy
Tucci et al.	1	Child	Nucleus	Common cavity	+	Canal wall down
Stattery and Luxford	2	Children	3M/House	Common cavity	+	None (facial recess)
			Nucleus		+	? Aberrant VII incus removed, Eustachian tube packed (facial recess)
McElveen et al.	3	Children	Clarion (2)	Common cavity	-	Labyrinthotomy
			Nucleus (1)	Common cavity	-	Labyrinthotomy

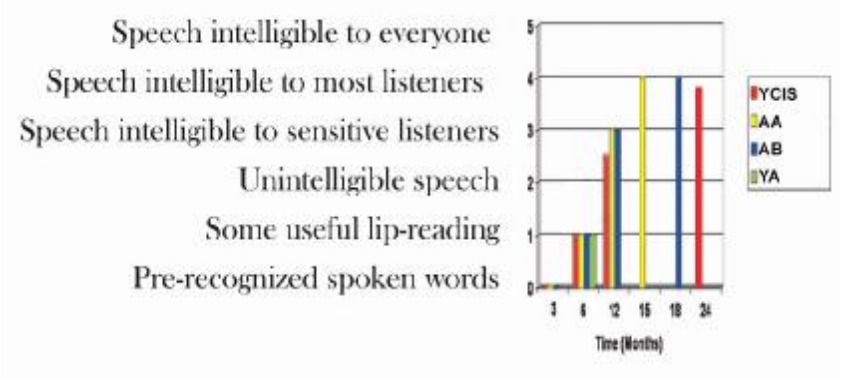
The alternative approaches that have been employed include the facial recess approach and the canal wall – down mastoidectomy with closure of the external auditory canal, although those alternative approaches may allow the surgeon access to the common cavity for placement of the electrode array, these approaches are associated with a higher rate of complications and may require unnecessary dissection. Using custom made devices prevents the electrode extending to the IAC. Tucci et al. (6), reported two patients with hypoplastic cochlear where the electrode array did extend into the IAC.

It is doubtful that these patients receive substantial vestibular input from the common cavity and instead rely more on visual and somatosensory information to maintain this equilibrium, consequently it is not surprising that despite accessing the common cavity through a location comparable to lateral semicircular canal, non of our patients experienced vertigo, a dizziness following cochlear implant, and this was also reported by McElveen.

After comparing our results regarding auditory performance and speech recognition over time, to those of YCIS (7), we concluded that after 18 months the results are surprisingly similar

A Comparison between Common Cavity and Normal Cochlea Cases (Regarding Auditory Performance and Speech Intelligibility)
Common cavity cases depending on our results
Normal cochlea cases depending on YCIS (Yorkshire Cochlea Implant Service), UK





Conclusion:
Common cavity deformities do not preclude successful cochlear implantation. speech discrimination and intelligibility was similar to implantation in patients with normal cochlea, as compared with the results of YCIS.

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