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Deaf hearing: Implicit discrimination of auditory content in a patient with mixed hearing loss

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ABSTRACT

We describe a patient LS, profoundly deaf in both ears from birth, with underdeveloped superior temporal gyri. Without hearing aids, LS displays no ability to detect sounds below a fixed threshold of 60 dBs, which classifies him as clinically deaf. Under these no-hearing-aid conditions, when presented with a forced-choice paradigm in which he is asked to consciously respond, he is unable to make above-chance judgments about the presence or location of sounds. However, he is able to make above-chance judgments about the content of sounds presented to him under forced-choice conditions. We demonstrated that LS has faint sensations from auditory stimuli, but questionable awareness of auditory content. LS thus has a form of type-2 deaf hearing with respect to auditory content. As in the case of a subject with acquired deafness and deaf hearing reported on a previous occasion, LS's condition of deaf hearing is akin in some respects to type-2 blindsight. As for the case of type 2 blindsight the case indicates that a form of conscious hearing can arise in the absence of a fully developed auditory cortex.

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1. Introduction

Some patients with neurological deficits show signs of implicit sensory detection, a phenomenon in which the subject may react in some way to a stimulus without

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being conscious of perceiving it (Weiskrantz, 1997, 1998a). In cases of blindsight, patients with damage to their primary visual cortex (V1) report being blind despite demonstrating appropriate responses to visual stimuli (Stoerig & Cowey, 1997).

The exact mechanism underlying blindsight is unknown, but there are data that suggest that in most cases of blindsight the retinal information is projected to subcortical structures that project directly to extrastriate regions, thus bypassing V1. It remains a possibility that some blindsight subjects have spared islands of V1 that carry information from subcortical structures to extrastriate regions. For example, one patient RD with extensive, unilateral lesions of V1 was found to have complete blindness in the right lower quadrant and residual vision in the right upper quadrant that probably was due to spared islands of functional tissue in V1 (Barbur, Watson, Frackowiak, & Zeki, 1993). However, the majority of blindsight subjects do not seem to have any spared islands in striate cortex that could explain the residual visual abilities (Zeki & Ffytche, 1998).

Although blindsight was originally defined as visual abilities in the absence of reported visual awareness, some subjects have been found to have conscious awareness in their affected hemifield despite extensive V1 lesions (Overgaard, 2011, 2012; Zeki & Ffytche, 1998). These patients appear to have residual vision for some stimulus attributes that they are unaware of, but they also show awareness of the presence and direction of fast-moving and/or high-contrast visual stimuli, and this awareness often positively correlates with their abilities to discriminate (Barbur et al., 1993; Zeki & Ffytche, 1998). The observation that some blindsight subjects have a form of residual awareness has resulted in a division of blindsight into type 1 and 2 (Weiskrantz, 1998a, 1998b). In type 1 blindsight, a subject with lesions to V1 has residual vision in the absence of reported awareness; in type-2 blindsight, subjects with lesions to V1 have a form of awareness that is positively correlated with their residual visual abilities. To what extent the phenomenology of type-2 blindsight is like degraded normal vision is still debated (Azzopardi & Cowey, 1997; Barbur, Weiskrantz, & Harlow, 1999; Brogaard, 2011a, 2012; Kentridge & Heywood, 1999; Overgaard, Fehl, Mouridsen, & Cleeremans, 2008; Overgaard & Grünbaum, 2011; Overgaard, Rote, Mouridsen, & Ramsøy, 2006; Sahraie et al., 1997; Weiskrantz, 2009). However, there is a growing consensus that type 1 and type 2 blindsight are manifestations of a single mechanism under different experimental conditions (Zeki & Ffytche, 1998; Brogaard, 2015). Given this hypothesis, both type 1 and type 2 blindsight might be able to shed light on the connection between the visual system and visual awareness.

At least one study has shown a possible auditory analog of type-2 blindsight: a patient with cerebral deafness displayed signs of largely unacknowledged detection of auditory stimuli (Garde & Cowey, 2000). The phenomenon is also known as *deaf hearing* (Cowey and Stoerig, 1992). Specifically, Garde and Cowey (2000) studied a patient with total deafness caused by a bilateral lesion in the temporal lobes and lesions in the central pontine area. The patient demonstrated some ability to respond reflexively to sounds, orienting her head appropriately in a

forced-choice paradigm, but she was unable to identify the meaning of the sounds presented to her.

Given the hypothesis that blindsight can provide insight into the connection between sensory areas and sensory awareness, a “model” of the phenomenon in a different modality is of great relevance. It potentially addresses whether blindsight illuminates something specific to the organization of visual areas in the brain, or whether it represents something more general about the organization of perceptual areas.

Not all people with functional deafness have a total inability to hear; instead, many have profound or severe hearing loss (Brookhouser, Worthington, & Kelly, 1990; Fraser, 1964). And even those with profound hearing loss may hear some frequencies better than others. It therefore makes more sense to talk about cortical deafness with respect to certain aspects of sounds, such as certain frequencies. Profound hearing loss is accompanied by deficits in perceiving amplitude – deafness may occur at some frequencies but not others. If deafness is understood in this way, deaf hearing does not require a total lack of auditory experience; rather it involves an ability to behaviorally detect or discriminate aspects of sounds that are not consciously perceived. That is, we predict that cases of deaf hearing may often be a form of type-2 deaf hearing. That is how we will use the term “deaf hearing” in this paper.

We studied a deaf individual, LS, whose hearing loss was discovered at the age of five. LS, now 54 years old, can hear most sounds with his hearing aid (see audiology chart below), but reports hearing nothing at all when not wearing his hearing aids. We designed five types of trials to determine whether LS could implicitly detect auditory stimuli that were explicitly undetected. We carried out a series of forced-choice paradigms to quantify sound detection, localization, and content discrimination. Further, we used structural magnetic resonance imaging (MRI) to quantify the features of LS’s auditory anatomy.

2. Methods and materials

All experiments were approved by the Institutional Review Boards at the University of Missouri, St. Louis and at Baylor College of Medicine. LS and control subjects gave written consent to participate in the study.

2.1. Subjects

Subject LS is a 54-year-old English-speaking man with 20 years of education and excellent abilities to speak and understand language. LS was born profoundly deaf in both ears but was not diagnosed until the age of five. After diagnosis, he was fitted with hearing aids and attended speech therapy and a regular elementary school. He was never taught sign language.

LS has both conductive and sensorineural loss (Figure 1). Conductive hearing loss is established by measuring the volume at which a sound must be played through the ear for it to be heard. LS's conductive hearing loss averages 95 dB, making it "profound." Sensorineural hearing loss is established by measuring the volume at which a sound must be played to the inner ear through the bones of the skull. LS's sensorineural hearing loss averages 75 dB, making it "severe." Mixed hearing loss is established by measuring the difference in decibels (dB) between conductive and sensorineural hearing loss. For LS, the difference averaged 15 dB, crossing the 10 dB threshold necessary to establish mixed hearing loss.

With-hearing-aids, LS is able to consciously hear middle- and low-frequency sounds. However, LS relies heavily on lip-reading and context to interpret spoken language, even with his hearing aids. LS reports that he is unable to hear anything without his hearing aids.

Like many other deaf individuals, LS suffers from tinnitus. However, he reports that he never pays much attention to it because he never experienced losing his sense of hearing. His tinnitus, he says, "is just background noise, much like the sound of air conditioning or cars on the street for hearing people."

LS started experiencing synesthesia when he was fitted with a hearing aid at the age of five. Synesthesia occurs when internal or external stimuli provoke atypical sensations or thoughts (Baron-Cohen, Wyke, & Binnie, 1987; Brogaard, Marlow, & Rice, 2014; Cytowic & Eagleman, 2009; Eagleman & Goodale, 2009; Grossenbacher & Lovelace, 2001; Hubbard, 2007; Hubbard, Arman, Ramachandran, & Boynton, 2005; Hubbard & Ramachandran, 2005; Ramachandran & Hubbard, 2001; Sperling, Prvulovic, Linden, Singer, & Stirn, 2005; Ward, Huckstep, & Tsakanikos, 2006). LS's synesthesia is polymodal, as he reports that vision (motion), touch, taste, smell, and emotion all give rise to an internal perception of sound. He reports that vision to sound is the strongest of the synesthetic associations. Additionally, he reports a bi-directionality with touch to sound synesthesia and sound to touch synesthesia.

LS's vision-sound synesthesia consists in a sound response to objects perceived in his environment. For example, LS experiences sounds corresponding to motion of objects in his visual field. LS reports that the vision-sound synesthetic associations exhibit automaticity and remain stable over time. For example, the sight of falling water droplets always gives rise to the sound of falling water droplets and the sight of a car passing by always gives rise to the same "whooshing" sound, even when the sound is not actually present.

Three hearing control subjects (2 females, 1 male; mean age = 44.6 ± 6) were involved in the detection, location, and content discrimination tasks. LS served as his own control when fitted with-hearing-aids.

2.2. Stimuli

Sounds were provided by the loop library in Apple's GarageBand software and played using Simple Soundboard installed on an Apple iMac. Given that audio

speakers may cause air vibrations that could provide cues as to a sound's presence or content, reference-grade Bose OE2 headphones were used for the sound detection, content discrimination, and perceptual awareness tests. However, given that LS's hearing loss differs by frequency in each ear, the isolation of sounds provided by headphones could interfere with the ability to localize the source of these sounds. Thus, for the source localization task, sounds were played over reference-grade speakers connected to a two-channel amplifier. Speakers were located 2 m to the left and right of the subject's head at a 45° angle. Three sounds were chosen for the tests, based on the uniqueness of their rhythms; these were "Band" (1.5 s), "Ringer" (1.5 s), and "Sonar" (1.5 s). Sound clips are available online at <http://brogaardlab.com/ls-sounds>.

2.3. Determining threshold

The methodology for each experiment was constructed to compare LS's ability to make correct judgments when wearing or not wearing his hearing aids. However, if headphones or speakers are loud enough, they can act as a substitute for hearing aids. To ensure that sounds were played below such a threshold for the without hearing aids condition, we used multiple ascending and descending sequences – we started very loud at above-threshold volume to which he reported hearing every time (without aids), and then we decreased the volume until he reported hearing zero times. We then started with the volume very low and below threshold and raised the volume to the point at which he reported hearing the sound every time. The determined thresholds were consistent in both directions. Sub-detection threshold output volume (OV) was set at approximately 10% less than its intended detection threshold (DT) (Headphones: 84.0 dB DT/75.6 dB OV for "Band," 102.0 dB DT/91.8 dB OV for "Ringer," and 81.0 dB DT/72.9 dB OV for "Sonar"; Speakers: 81.0 dB DT/72.9 dB OV for "Band," 98.0 dB DT/88.2 dB OV for "Ringer," and 81.0 dB DT/ 72.9 dB OV for "Sonar"). All sounds played to LS during the without hearing aids condition were played at these sub-threshold output volumes. For the with-hearing-aids condition for LS and all conditions for controls, the output volumes of the sounds were adjusted to a comfortable level, maintaining the relative difference in decibels among the sounds (Headphones: 61.0 dB OV for "Band," 68.0 dB OV for "Ringer," and 62.0 dB OV for "Sonar"; Speakers: 68.0 dB OV for "Band," 82.0 dB OV for "Ringer," and 66.0 dB OV for "Sonar"). Output volume was calibrated using a digital sound level meter at a distance of 2.0 m from each speaker and a distance of 2.0 cm from each headphone. Sub-detection threshold was verified before each trial.

2.4. Experimental design

2.4.1. Trial setup

For LS, each trial used a forced-choice paradigm to test his judgment in the with and without hearing aids conditions. When LS could not hear a sound, he was

forced to guess. All sounds in the without hearing aids condition were played at sub-threshold volume. LS was placed in a soundproof, darkened room and blindfolded to avoid any auditory interference from his vision-sound synesthesia.

2.4.2. Task 1: Sound detection

The first task determined whether LS could detect the presence of sounds that he denied hearing. LS was presented in random order either with one of the sounds playing repeatedly on a loop or with silence. After 5 s, LS was tapped on the neck, indicating that he should raise his left hand if he felt a sound was present or his right hand if no sound was present. This process was repeated with 8 s of silence between trials. LS completed 10 trials for each of the three sounds for a total of 30 trials, and 30 more randomly interleaved trials in which no sounds were played.

2.4.3. Task 2: Source localization

We next determined whether LS was able to localize the source of sounds that he denied hearing. LS was presented in random order with sounds playing repeatedly on a loop 1 m to his left or right. After 5 s, LS was tapped on the neck indicating he should raise his left or right hand to indicate from which direction the sound originated. LS completed 30 trials, totaling 5 trials for each of the sounds played to the left or right ear.

2.4.4. Task 3: Content discrimination

The third task determined whether LS could discriminate the content of sounds that he denied hearing. LS was placed in a soundproof, darkened room with his hearing aids on and blindfolded to ensure he did not gather any visual cues from the experimenter. We used a forced-choice paradigm similar to the one used to test for location detection. This task was split into three parts, beginning with the with-hearing-aids condition, which served as a control measure and allowed LS to become familiar with the sounds that he would be asked to discriminate. At the beginning of each sound trial, one of the three sounds or no sound was played in random order repeatedly on a loop. The recorder was blind as to the condition. After 5 s, LS was tapped on the back of the neck indicating he should raise his right hand to indicate what sound was being played: a boxing move represented “band,” a circulating finger represented “ringer,” a talking hand movement represented “sonar,” and a flat hand represented “no sound.” He was also instructed to point his left thumb up or down to indicate whether he heard a sound. This process was repeated with 8 s of silence between trials. LS completed 80 trials, totaling 20 trials for each of the sounds and silence. Each stimulus occurred with equal probability.

2.4.5. Task 4: Perceptual awareness with and without training

Task 4 determined the extent of LS’s awareness of auditory stimuli by using an auditory equivalent of Overgaard and colleagues’ *Perceptual Awareness Scale* (PAS), a four-point scale with the following possible answers: “not seen” (NS); “weak

glimpse” (WG); “almost clear image” (ACI); and “clear image” (CI) (Overgaard et al., 2006, 2008). An auditory version of PAS was developed in a previous experiment, directly comparing visual and auditory performance and report, showing a highly similar tendency (Overgaard et al., 2013). The scale does not just represent four arbitrary points; the points have a specific meaning. Thus, NS represents a case in which not even the faintest sensation is experienced when a subject is presented with a stimulus. Subjects select WG when they have some weak experience without the ability to specify the content hereof. For ACI and CI, the content can be specified but with different degrees of clarity and certainty (See Sandberg & Overgaard, 2015 for a detailed description of the methodology).

The scale was developed in collaboration with subjects during long training sessions (Ramsøy & Overgaard, 2004), and directly compared with other measures of conscious experience (Sandberg et al., 2010), and with binary reports (Overgaard et al., 2006, 2008), revealing that not only is PAS arguably more sensitive than other measures but that healthy as well as brain injured participants classify individual experiences differently. Using a binary measure, participants classify experiences as “not seen/heard” which they, using PAS, may classify as “weak glimpse.”

For this task, we repeated Task 3 with one crucial change: LS was asked to indicate his awareness of the sound during each trial by raising between one and four fingers on his left hand. One finger indicated that the sound was “not heard” (NH), two fingers indicated a “weak glimpse” (WG), three fingers indicated an “almost clear sound” (AC), and four fingers indicated a “clear sound” (CS). Similar to Overgaard and colleagues (2008), we divided the PAS task into two parts in order to measure how a more detailed understanding of the awareness scale might lead to a stronger correlation between reported awareness and success in discriminating among content. For the first part of this trial, LS was asked to use the PAS scale without clarification of what each point meant. After this first part, we interviewed LS to understand how he assigned different PAS points to his perceptual experience. LS indicated that the pattern of his tinnitus became more distinct, giving him greater confidence in his choice of which sound was playing. In these cases, LS would report a higher awareness of the sound using the PAS scale, despite not actually hearing the sound.

Based on this PAS interview, we then gave LS the conditions that must be met for each PAS point. For the second part of this trial, LS was instructed to use the NH condition when he could not hear any sound and had no “gut feeling” corresponding to his answer. In other words, NH was reserved for when he felt he was making a random guess. LS was instructed to use the WG condition when he could not hear any sound, but he had a gut feeling corresponding to his answer. The AC condition was reserved for when LS could not hear the sound but was able to answer based upon the change in characteristics of his tinnitus. Finally, LS was instructed to use CI when he heard the sound. LS completed 80 trials each for parts 1 and 2, totaling 20 trials for each of the sounds and silence. LS was not aware that only three-fourth of the trials contained sounds.

2.4.6. MRI data acquisition

In order to characterize the vision-sound synesthesia reported by LS, structural MRI studies were completed at Baylor College of Medicine. MRI data were acquired using a 3 Tesla Siemens Trio scanner and a Siemens 12-element head coil at Baylor College of Medicine's Center for Advanced MRI. An anatomical MPRAGE scan was acquired for the purpose of aligning the functional data (T1-weighted, TR = 1200 ms, TE = 2.66 ms, 245 mm FOV, 1 mm slice thickness). The EPI-BOLD sequence (37 slices, TR = 2000 ms, TE = 25 ms, flip angle = 90 degrees, 64 x 64 matrix, 3.4 x 3.4 mm voxel size, 4 mm slice thickness) was acquired after the MPRAGE.

Given the unique character of LS's reports in regard to his synesthesia, it was unlikely that the utility of acquiring novel control data would outweigh the costs of data acquisition, thus control data was not collected at the time of this study.

FreeSurfer (v5.0; <http://surfer.nmr.mgh.harvard.edu/>) was used to calculate raw regional gray and white matter from LS's high-resolution T1-weighted structural MRI images.

We used Crawford and Howell's (1998) modified t test to compare LS's brain measurements to hearing and deaf control subjects taken from Emmorey, Allen, Bruss, Schenker, and Damasio (2003). For the a priori region of interest (ROI) (gray matter: superior temporal gyrus), we set statistical significance at $p < .05$, one-tailed. We hypothesized that LS's gray matter volume in this ROI would be smaller than the corresponding ROI in the hearing and deaf controls.

3. Results

3.1. Task 1: Sound detection

The aim of this task was to determine whether LS could detect the presence of sounds that he denied hearing when he was not wearing hearing aids. On subthreshold stimuli, LS insisted that he did not hear the sounds, but the task forced him to report whether or not a sound was present. LS was able to report all 30 instances of sounds played when he was wearing aids as well as report all 30 instances of sounds played above the detection threshold when he was not wearing aids. However, LS correctly reported only 17 out of 30 instances of sounds playing and 14 out of 30 instances of silence, for a total of 31 correct identifications out of 60 when he was not wearing his hearing aids. A Fisher exact probability test was used to test whether or not the proportion correct when sounds playing ($M = .567$) was significantly different than the proportion correct when there is no sound ($M = .495$) and it indicated that he does not have deaf hearing with respect to sound detection, $p = .606$.

3.2. Task 2: Source localization

After it was determined that LS, without his hearing aids, could not identify when a sound was played, the next task was designed to understand whether LS could

reliably localize the sound source (to the right or left). For the without hearing aids condition, LS correctly localized sounds in 14 out of 30 of the trials, indicating he scored no better than chance. For the with-hearing-aids condition, LS correctly localized sounds in 23 out of 30 trials. A Fisher exact probability test was used to test whether or not the proportion correct with-hearing-aids ($M = .767$) was significantly different than the proportion correct without hearing aids ($M = .467$), and indicated a significant difference between two conditions, $p = .033$. Three hearing control subjects performed at ceiling (Figure 2).

3.3. Task 3: Content discrimination

We then tested how LS would perform if asked to determine the content rather than to localize the sounds. LS was asked to discriminate among sounds using a unique hand signal for each sound, and a separate signal when he heard no sound at all. Overall, LS correctly identified which sound was playing or if silence was playing on 56 trials out of 80 ($M = .7$) without hearing aids. Chance level for this experiment was defined as .25, given that there are four options. A binomial test was conducted to test if the proportion correct without hearing aids ($M = .7$) was significantly different than the chance level, and the binomial test indicated a significant difference, $p < .001$. When the proportion of correct decisions was examined by the type of sound, LS correctly identified 9 ($M = .45$), 18 ($M = .90$), 14 ($M = .70$), and 15 ($M = .75$) trials out of 20 for Band, Ringer, Sonar, and No Sound cases, respectively. A binomial test indicated a significant difference than chance success for all types of sounds at the alpha level of .05. In the discussion, we will return to the surprising finding that he often made the correct hand signal for silence, even though he performed at chance in the Sound Detection task (Task 1) (Figure 3).

3.4. Task 4: Perceptual awareness with and without training

To determine the extent of LS's awareness of auditory stimuli, we used an auditory equivalent of Overgaard and colleagues' PAS, which was originally created specifically for visual stimuli. In the without hearing aids condition, LS reported no instances of hearing a "clear sound" (CS), indicating that he never heard the sound clearly. Before PAS training, LS only answered "not heard" (NH) and "weak glimpse" (WG). The proportion of correct responses was .81 (59 correct out of 73 trials) when there was no instruction, and was .53 (29 correct out of 55 trials) after instruction for the NH condition. Due to the large sample size, Fisher exact test was computationally not feasible, so a chi-square test was conducted and indicated a significant relationship between instruction and proportion of correct responses for the NH condition, $p < .01$. Proportion of correct responses for NH condition significantly decreased after instruction. The proportion of correct responses was .86 (6 correct out of 7 trials) when there is no instruction,

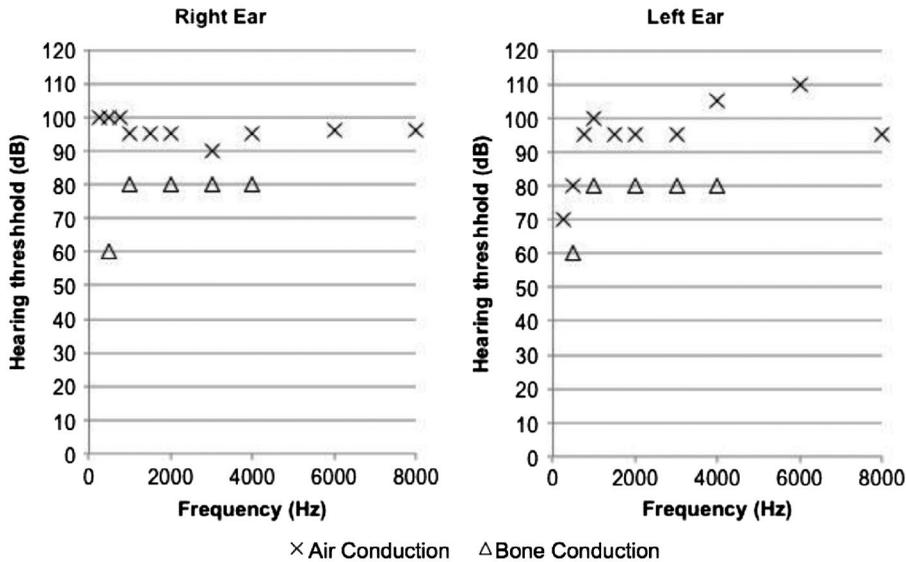


Figure 1. Audiometry report for LS taken 09 September 2011. Profound hearing loss is defined as greater than 90 dB of hearing loss in the better ear. LS exhibits profound hearing loss. LS experiences middle-pitched sounds only at very high amplitudes.

and was .84 (16 correct out of 19 trials) after instruction for the WG condition. A Fisher exact test indicated there was not a significant relationship between instruction and proportion of correct responses for WG condition, $p = .99$. For the AC condition, the proportion of correct response was 100% (4 correct out of 4 trials) after instruction.

Overall, LS showed no significant difference in ability to discriminate among content when offering PAS reports rather than when offering binary (yes/no) reports ($p = .319$) (Figure 5).

These results are as predicted from previous experiments with visual stimuli (Sandberg, et al., 2010), with auditory stimuli (Overgaard et al., 2013), and with a blindsight patient (Overgaard et al., 2008).

3.5. MRI

In the functional MRI experiment, our goal was to probe LS's motion-sound synesthesia. We contrasted blocks of moving dots with static dots to see whether the synesthetic sound could be evidenced by auditory cortex activity. LS reported auditory synesthetic experiences during motion blocks, and the results indeed revealed a significant cluster in right auditory cortex induced by visual motion (Figure 6).

The structural MRI scan revealed no clear structural damage to the auditory cortex (Figure 7). However, the volume of LS's bilateral superior temporal gyri (STG) – 12.6 and 11.5 cm³ for left and right hemispheres, respectively – are smaller

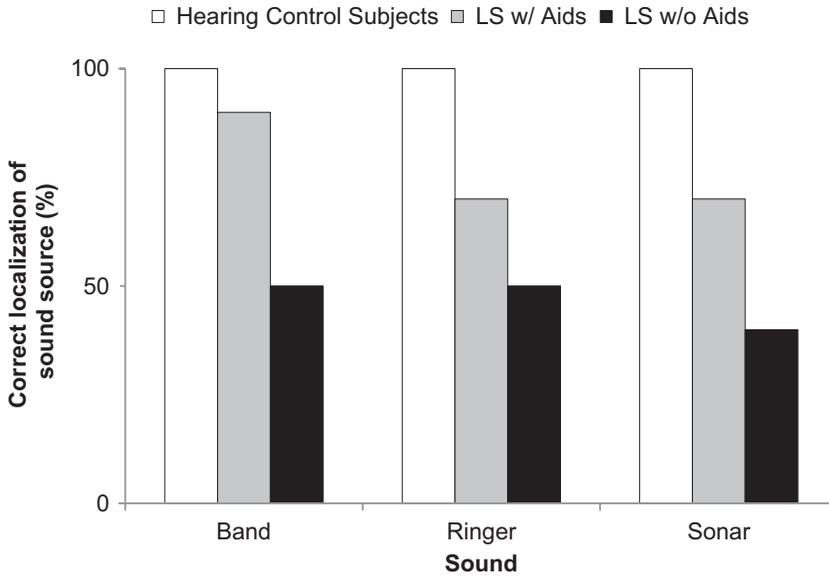


Figure 2. LS cannot localize sounds without his hearing aids (Task 2). The Y-axis represents the number of correct localizations (right/left) of one of the sounds “band,” “ringer,” or “sonar.” The number of maximum correct answers was 30. The hearing control subjects ($n = 3$) scored correctly on all sound clips. Where chance is defined as 50% correct answers, LS’s guesses were not significantly above chance when he was not wearing his hearing aids. LS’s guesses were also not significantly different than chance when he was wearing his hearing aids. What is shown is whether or not there is a difference in proportion of corrects in two different conditions. The fisher exact test indicates they are significantly different as reported.

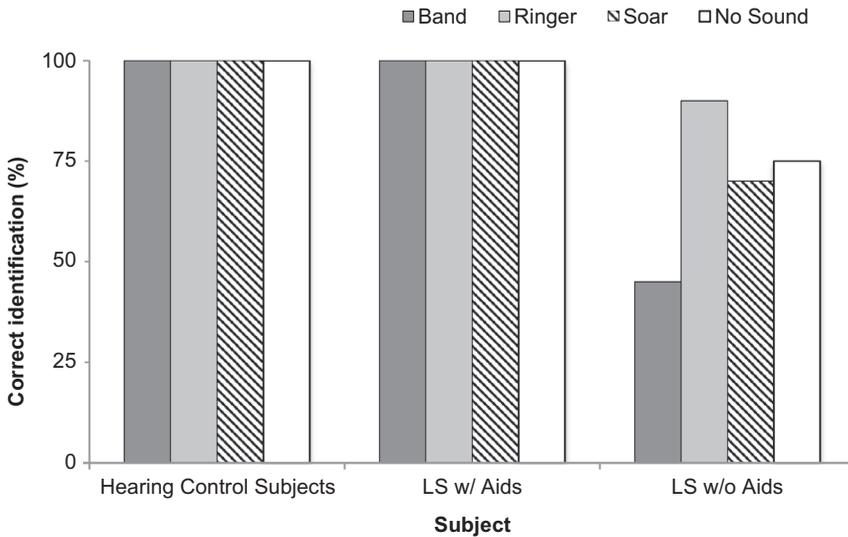


Figure 3. Even without hearing aids, LS can identify sounds above chance. The Y-axis represents the number of correct identifications of one of the sounds “band,” “ringer,” or “sonar” or no sound played. The maximum correct answers were 20 for each condition. Where chance is defined as 25% correct answers, LS’s guesses were significantly above chance even when he was not wearing his hearing aids.

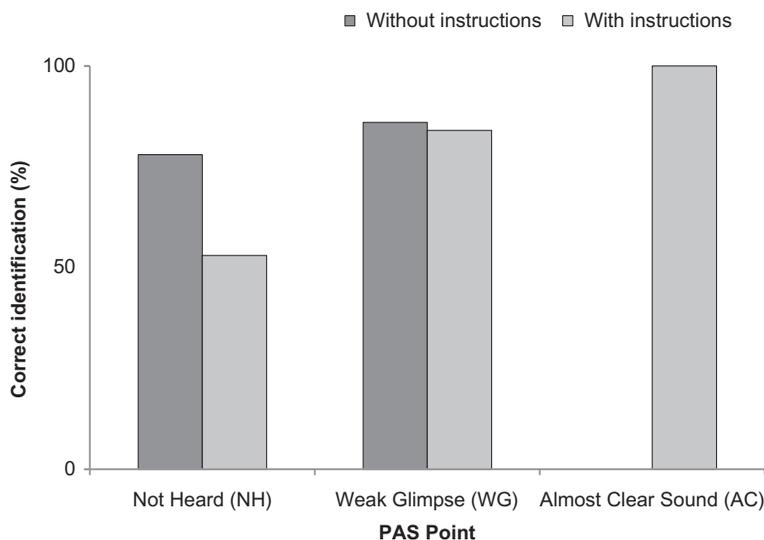


Figure 4. Results of the Perceptual Awareness Task. The PAS charts represent the probability that LS answered correctly given the strength of his experience (NH, WG, etc.). Each bar is calculated by dividing the number of correct answers for each point on the scale by the total number of times LS used that point. The Y-axis represents LS's correct identification of one of the sounds "band," "ringer," or "sonar." Fully correct identification was 1 and was achieved only when LS reported having an "almost clear" image.

than the average for hearing controls (16.1 ± 3 and 13.8 ± 3.0 cm³; Emmorey et al., 2003) and smaller than for congenitally deaf subjects (16.0 ± 2.8 and 15.2 ± 2.8 cm³, data from Emmorey et al., 2003; see Tables 1–3).

4. Discussion

We carried out several tasks to test LS's abilities to detect, localize and identify the content of auditory stimuli that he reported being unable to consciously hear without hearing aids. For all trials, LS was tested with and without hearing aids. On the location test, LS performed comparably to three controls when allowed to use aids. However, in the without hearing aids condition, LS performed at chance in the sound detection and source localization trials. LS thus does not have "unconscious hearing" with respect to the determination of the presence and location of sounds.

However, when we probed LS's ability to discriminate among auditory content in the without hearing aids condition, LS's judgments were above chance for all three sounds and for a silence stimulus. LS thus appears to the ability to report *auditory content*. Because of the forced-choice nature of the test and the non-conscious nature of the ability, LS was unable to rely on the auditory content to correctly determine the presence and location of sounds in Tasks 1 and 2.

We also investigated the extent to which LS was aware of sounds in the without hearing aids condition. Using a variation on Overgaard and colleagues' PAS

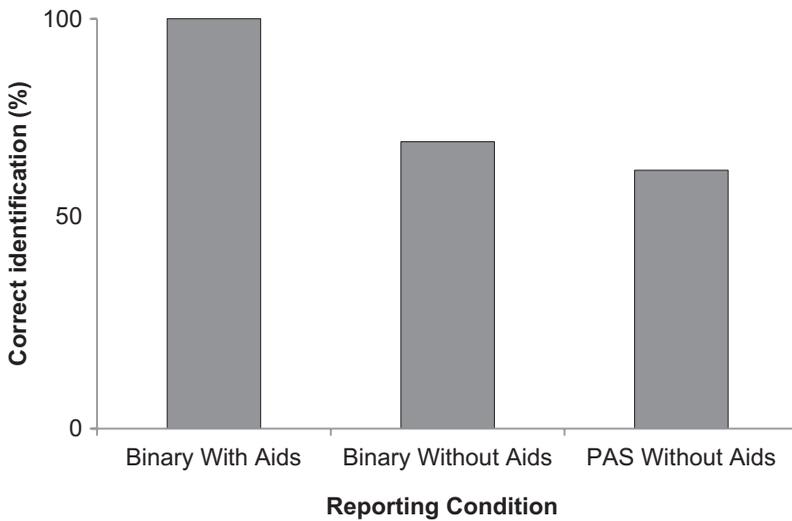


Figure 5. Comparison of Binary and PAS Results: The Y-axis represents LS's correct identification of one of the sounds "band," "ringer," "sonar," or no sound. There was no significant difference in LS's ability to discriminate among content when he was asked to provide a PAS report rather than indicate whether the sound was heard or unheard (binary report.).

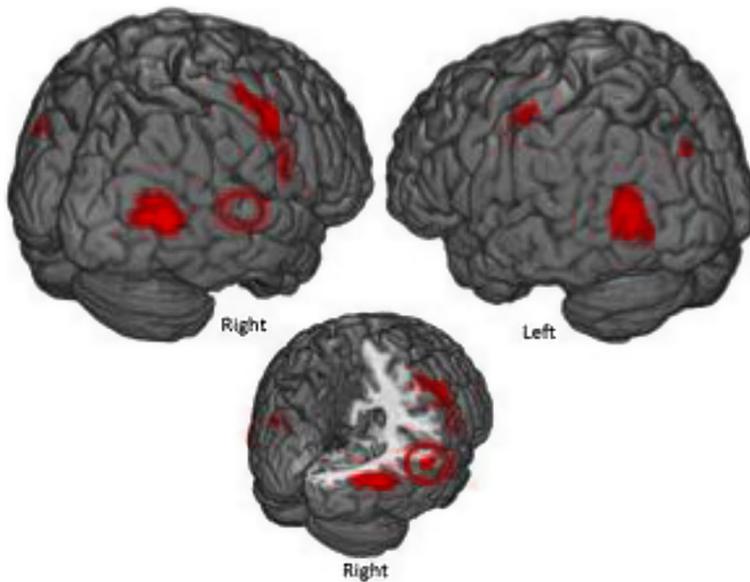


Figure 6. When LS sees moving dots, functional MRI results show activity in bilateral MT (expected motion activity), pre- and postcentral gyri, and a 63-voxel cluster in the right superior temporal gyrus (circled in red).

scale, it was determined that LS's ability to make correct judgments of auditory content correlated with the reported degree of awareness (Figure 4). However, there was no significant difference in LS's ability to discriminate among auditory

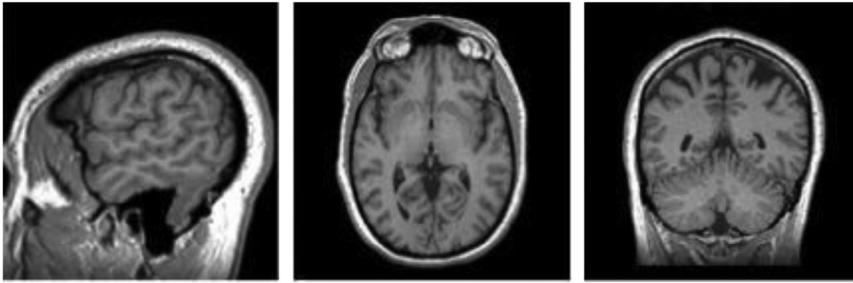


Figure 7. (a) Sagittal ($x: -50.24$ mm; $y: -50.07$ mm; $z: 26.60$ mm), (b) axial ($x: -98.88$ mm; $y: -88.02$ mm; $z: -7.5$ mm) and (c) coronal views ($x: 102.46$ mm; $y: 27.28$ mm; $z: 97$ mm). The volumetric assessment (Table 2) shows that LS's right and left STG are significantly smaller than the mean for congenitally deaf subjects (11.5 and 12.6 cm³ vs. 15.2 and 16.0 cm³; $SD = 2.8$). The means are from Emmorey and colleagues (2003).

content in the without hearing aids condition when also asked to report on his level of awareness based on a four-point rather than binary scale (Figure 5). More speculatively, perhaps the tinnitus serves as a mask – blocking off conscious access to detection, but not interrupting implicit links to an object recognition system. That is, it may have interfered with the “where” system, but less so with the “what” system. The results here would then be more similar to work on masking and implicit decisions than the blindsight literature.

The mechanism underlying deaf hearing is unknown. It is plausible that LS's faint auditory awareness played a role in his ability to discriminate auditory content. From such a perspective, LS's particular brain organization (a smaller than expected superior temporal gyrus) would allow information to be processed to a certain level of representation associated with weak auditory experience (Kouider et al., 2010; Overgaard & Mogensen, 2011).

The fact that LS was able to discriminate among auditory content even though he could not detect or localize sounds is indeed a surprising result. This is the opposite result of the patient in the Garde and Cowey (2000) study, wherein the deaf-hearing subject demonstrated the ability to detect and localize sounds but was unable to discriminate among auditory content, which may indicate a different pathway for deaf hearing in each patient.

Furthermore, responding to meaning appears to require not only determining low-level features of auditory content (e.g., pattern, pitch, and timbre), but also determining high-level features of auditory content (e.g., band playing, ringing, sonar sound). Given that LS denied even hearing the sounds presented to him, determining high-level features of auditory content seems to be an unusually complicated task.

The surprising results of the current study may be explained by the different nature of the tasks. Although discrimination among the auditory content of sounds may, at first, seem more difficult than the determination of the presence or location of sounds, there are some reasons to think that the former task requires less of the

auditory system than the latter. LS's ability to discriminate among content may be facilitated by a mixture of auditory and non-auditory cues relating to the high-level properties composing the auditory content. LS may be using his highly developed language-processing center together with faint signals in auditory cortex to complete the meaning task successfully. Thus, whereas the auditory processing of the stimulus may not have sufficed for LS to gain access to the processed information, it is plausible that the auditory processing in tandem with semantic processing (e.g., through top-down influences) led to a stronger signal that he could access in a forced-choice paradigm.

Sound localization may be particularly difficult for LS, due to his profound conductive hearing loss. Locating a sound to the right or left requires numerous auditory cues, including interaural time differences (sound from the left reaches the left ear earlier than the right ear because the head blocks the path of the sound) together with phase delays in the case of low frequencies and group delays in the case of high frequencies (Blauert, 1983). LS's conductive hearing loss may have impeded the chance of picking up on cues to sound location coming from interaural time differences.

The structural MRI scan supports the hypothesis that the signals in auditory cortex may have been too weak for LS to access cognitively unless strengthened by top-down influences, as in the content discrimination task. According to the volumetric data (Tables 1 and 2), LS's STG are significantly smaller than those of congenitally deaf individuals and his left superior temporal gyrus is significantly smaller than that of hearing controls. The STG are not normally smaller in congenitally deaf people than in hearing subjects, because these areas typically take on new functions (Shibata & Kwok, 2006). Visual or multimodal inputs from the occipital and parietal cortex may replace auditory inputs from regular auditory pathways (Fine et al., 2005; Finney et al., 2001; Meredith & Allman, 2012; Penhune et al., 1996, 2003; Shibata, 2007). If we take the volumetric data at face value, the relatively small size of LS's STG may indicate that LS has functional defects in the auditory cortex, perhaps due to an underdeveloped or incomplete tone mapping. Brain reorganization, if it has occurred, as well as processing of sounds from the hearing aids may not have given rise to a gray matter increase as is normally seen in congenitally deaf individuals. If the auditory cortex is underdeveloped, this may lead to the inability to perform sound detection and localization tasks successfully. However, faint signals from the auditory cortex may be strengthened by feedback from the language-processing center allowing for successful content discrimination.

LS remained certain that he had not heard any sounds during the without hearing aids condition, regardless of his response. One explanation of this reported lack of auditory awareness may be that the content of his auditory experience lacked the auditory information necessary for full auditory awareness. Patients with both forms of blindsight have cognitive access to wavelength information but lack access to brightness information (Brent et al., 1994; Morland et al., 1999; Stoerig

& Cowey, 1989, 1992). It has been argued that although wavelength information allows blindsighters to make above-change judgments under forced choice, both wavelength and brightness information are necessary for full visual consciousness (Brogaard, 2011a, 2012, 2015). Likewise, LS's judgments may be based on information that is too restricted to provide for genuine auditory experience.

An alternative and more speculative explanation of LS's lack of full auditory experience associated with auditory content discrimination and his insistence on not hearing the stimuli is that he does not use the regular ventral stream auditory pathway to process auditory information. The processing of auditory content is in some ways analogous to the processing of visual content. According to the two streams hypothesis, visual information is processed in two cognitive streams that split from the primary visual cortex (V1). The dorsal visual stream runs upward through the parietal cortex, while the ventral visual stream runs sideways through the temporal lobe. The ventral visual stream is specialized for continuing vision for action whereas the dorsal visual stream is specialized for vision for object recognition (Goodale & Milner, 1992; Milner & Goodale, 1996, 2008). Although the visual dorsal stream is capable of performing complex calculations necessary to direct action, dorsal stream processes associated with vision for action do not produce reportable visual awareness (Brogaard, 2011a, 2011b; Milner & Goodale, 1996).

Much like in the visual system, there is segregated processing of auditory information in anatomically and functionally separate ventral and dorsal processing streams (Cisek & Turgeon, 1999; Rauschecker & Scott, 2009; Wang et al., 2008). "What" information and "where" information are separated in the auditory system in a manner analogous to the visual system. With respect to audition, the "what" system is involved in using auditory information to identify a sound. The "what" system forms the basis of both speech perception and music perception. The "what" system starts in the core region and then moves to more anterior parts of the temporal lobe. The "where" system is responsible for localizing sound in space. The "where" system also begins in the core region of the auditory cortex and then moves to posterior regions of the temporal lobe, as well as the posterior parietal cortex (Rauschecker & Scott, 2009).

It is plausible that LS was relying on the dorsal auditory pathway for auditory content discrimination in the forced-choice paradigm. In all other known cases of deaf hearing, participants sustained damage to previously functioning auditory systems. LS's deaf hearing, however, likely was present during early development. At that time, his brain could not rely on auditory information to detect and localize potential action-guiding stimuli, so the particular areas of the auditory dorsal stream that normally process action-guiding auditory information were rendered useless. Because of a diminished capacity to utilize audition for action, the dorsal auditory pathway might have undergone reorganization to process auditory information that LS can access in forced-choice paradigms. Dorsal stream representations in the vision-for-action pathway are not normally associated with reportable

sensory consciousness (Brogaard, 2011a, 2011b; Goodale & Milner, 1992; Milner & Goodale, 1996, 2008). If the same is true for dorsal stream representations in the audition-for-action pathway, this would explain the reported lack of auditory consciousness associated with successful auditory content discrimination.

One lesson about sensory awareness that is possible to draw from the study is that auditory consciousness appears to be possible in the absence of a fully developed superior temporal gyrus. A similar hypothesis has previously been put forward about type-2 blindsight, supported by different findings (Barbur et al., 1993; Ffytche & Zeki, 2011; Overgaard, 2012; Weiskrantz, Barbur, & Sahraie, 1995; Zeki & Ffytche, 1998). The widely studied blindsight subject GY was found to have large lesions to striate cortex that could not fully account for his residual visual abilities or the residual awareness under high contrast/high speed conditions (Barbur et al., 1993; Weiskrantz et al., 1995). It is widely thought that GY's residual vision in the absence of awareness involves direct projections from subcortical areas to extrastriate regions. Using an fMRI paradigm, Zeki and Ffytche (1998) found that both fast-moving stimuli associated with awareness and slow-moving stimuli not associated with awareness in GY led to activity in V5/MT but at different levels of intensity. They also found covariation in the dorsal stream (area V3 and parietal cortex) as well as the right middle frontal gyrus, but it is unclear to what extent this activity contributed to GY's residual awareness and visual abilities. On the basis of the data from studies of GY and other subjects with both type 1 and type 2 blindsight, Zeki and Ffytche (1998) hypothesized that the two conditions are manifestations of a single mechanism under different conditions. V5/MT receives its input directly from V1 (Cragg, 1969) but there are projections to this area directly from the lateral geniculate nucleus (Benevento & Yoshida, 1981; Fries, 1981; Yukie & Iwai, 1981) and via the superior colliculus projecting to the pulvinar nucleus, which in turn projects to V1 (Benevento & Standage, 1983). Thus, a likely mechanism underlying type 1 and type 2 blindsight is that direct projections to V5/MT from subcortical regions bypassing V1 can result in vague conscious awareness or residual vision without awareness depending on the contrast and speed of the stimulus. It therefore appears that some forms of conscious vision can arise in the absence of functional striate cortical areas. Likewise, the case of LS seems to indicate that some form of conscious hearing can arise in the absence of a fully developed auditory cortex.

5. Conclusion

We have described a patient LS who is profoundly deaf in both ears, possibly owing to underdeveloped temporal lobes from birth. Without his hearing aids he was unable to make above-chance judgments about the presence and location of sounds when presented with a forced-choice paradigm. However, he was able to discriminate high-level auditory content when forced to identify *which* of three sounds (or a silence) was presented. A perceptual awareness test indicated that

he had weak experiences of sounds, which may have assisted in his successful auditory content discrimination without his hearing aids. However, LS's weak experiences of sounds do not explain the discrepancy in LS's ability to discriminate auditory content while being able to detect the presence or location of sounds. We hypothesize that the auditory cues necessary for sound detection and localization did not suffice for LS to access the information in the forced choice paradigm. With respect to auditory content, there may be more cues to draw upon. This may have allowed LS successfully to discriminate content under forced choice. LS's lack of full auditory consciousness associated with successful content discrimination may be explained by the hypothesis that the part of his auditory system involved in audition for perception is defective, leaving the signals in the auditory system too faint to be fully consciously detected. Given that LS likely is relying on an alternative auditory pathway to process auditory information, we hypothesize that conscious hearing can arise in the absence of a fully developed auditory system.

Disclosure statement

No potential conflict of interest was reported by the authors.

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Appendix.**Table 1.** Volumetric data for subject LS. All data are in mm³.

Location	Right volume (mm ³)	Left volume (mm ³)
<i>Temporal lobe</i>		
Posterior bank	2164	2298
Posterior bank white matter	2808	2828
Entorhinal cortex	1636	2229
Entorhinal white matter	487	904
Middle temporal gyrus (MT)	10908	11601
Middle temporal white matter	5358	5336
Superior temporal gyrus	11395	12340
Superior temporal white matter	6776	7773
Temporal pole	2483	2439
Temporal pole white matter	643	581
Heschel's gyrus	953	1262
Heschel's gyrus white matter	571	863
Parahippocampal gyrus	2558	2775
Parahippocampal white matter	2006	1742
<i>Frontal lobe</i>		
Caudal middle frontal gyrus	5233	5111
CMF white matter	5366	6591
Paracentral lobule	2598	2261
Paracentral white matter	3244	2826
Pars opercularis	3621	4364
Pars opercularis white matter	3467	4068
Pars orbitalis	2723	1865
Pars orbitalis white matter	1345	870
Pars triangularis	4394	4691
Pars triangularis white matter	3651	4168
Precentral gyrus	11089	11606
Precentral white matter	13199	14376
Rostral middle frontal gyrus	14780	15131
RMF white matter	13117	12938
Superior frontal gyrus	18959	20992
Superior frontal gyrus matter	17340	18713
Frontal pole	798	792
Frontal pole white matter	267	336
<i>Parietal lobe</i>		
Inferior parietal lobule	14428	11104
Inferior parietal white matter	11317	9569
Pericalcarine cortex	2223	1718
Pericalcarine white matter	2966	3662
Postcentral gyrus	7829	8240
Postcentral white matter	8058	7665
Precuneus	9123	9460
Precuneus white matter	9587	9696
Superior parietal lobule	13581	12344
Superior parietal white matter	12022	13708
Supramarginal gyrus	10784	11743
Supramarginal white matter	9455	9100
<i>Occipital lobe</i>		
Cuneus (V1)	2544	2437
Cuneus (V1) white matter	2109	2617
Lateral occipital cortex	13917	12668
Lateral occipital white matter	11863	11076
Lingual gyrus	7093	5825
Lingual white matter	5643	5466
<i>Cerebellum</i>		
White matter	16349	17085
Cortex	62166	59413

Table 2. Structural analysis of LS's gray matter brain volume indicate that he may have underdeveloped temporal lobes STG = superior temporal gyrus. The average sizes for hearing and deaf individuals for STG are from Emmorey and colleagues (2003).

Sector	Hearing mean (SD)	Deaf mean (SD)	LS	t-test	
				p-value (hear)	p-value (deaf)
Left STG	16.1 (3.6)	16.0 (2.8)	12.6	<.05	<.05
Right STG	13.8 (3.0)	15.2 (2.8)	11.5	ns	<.05

Table 3. Significant cluster coordinates from the contrast of moving stimuli over static stimuli in patient LS.

Coordinates (MNI)	Cluster size	T-value
52 -70 12	625	6.11
-46 -76 12	744	5.51
56 -12 48	319	4.13
60 -32 6	63	3.81
62 -10 22	70	3.58
-18 -88 38	27	3.40
-54 -22 52	35	3.23