**Supplemental Table 1.** Summary of all included articles.

|  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- |
| Author | Year | Country | Study Type | Number of Implants | Pathology | Gender (%M) | Age (SD) |
| Alshehabi15 | 2019 | Ireland | retrospective chart review | 8 | SS | 0.75 | 49.875 (15.9) |
| Amoodi16 | 2012 | Canada | case report | 2 | VS | 0.50 | 42.5 (23.3) |
| Artukarslan17 | 2021 | Germany | retrospective chart review | 6 | SS | 0.50 | 62.8 (7.3) |
| Berrettini18 | 2012 | Italy | case report | 3 | SS | 1.00 | 59 (20.1) |
| Bird19 | 2010 | New Zealand | letter to editor | 1 | SS | 1.00 | 56 (NA) |
| Bittencourt20 | 2012 | Brazil | retrospective case report | 2 | SS | 1.00 | 50.5 (16.3) |
| Carlson21 Study **1** | 2012 | U.S. | retrospective case series | 5 | VS | 0.60 | 44.8 (11.3) |
| Carlson22 Study **2** | 2016 | U.S. | retrospective case review | 10 | VS | 1.00 | 63 (16.8) |
| Clark75 | 2001 | U.K. | case report | 1 | Brain or skull base radiation | 0.00 | 34 (NA) |
| Costello24 | 2016 | U.S. | case report | 1 | VS | 0.00 | 68 (NA) |
| Deep5 | 2021 | U.S. | retrospective case review | 12 | VS | 0.25 | 51.4 (16.5) |
| Ebode25 | 2021 | France | retrospective case series | 6 | Neurosarcoidosis | 0.67 | 36.8 (10.8) |
| Eitutis8 | 2021 | U.K. | retrospective chart review | 8 | VS | 0.00 | 48.8 (15.1) |
| Formanek26 | 1998 | Austria | case report | 1 | Brain or skull base radiation | 1.00 | 67 (NA) |
| Fujimoto27 | 2007 | Japan | case report | 1 | SS | 1.00 | 57 (NA) |
| Greene28 | 2017 | U.S. | case report | 1 | Neurosarcoidosis | 1.00 | 39 (NA) |
| Grover29 | 2011 | U.K. | case report | 2 | SS | 1.00 | 55 (1.4) |
| Halyur30 | 2021 | India | case report | 1 | VS | 0.00 | 32 (NA) |
| Harris10 | 2017 | U.K. | retrospective case review | 9 | VS | 0.44 | 48.7 (13.5) |
| Hathaway31 | 2006 | U.S. | retrospective case review | 1 | SS | 0.00 | 44 (NA) |
| Häußler32 | 2021 | Germany | retrospective case series | 3 | VS | 0.67 | 66.7 (11.0) |
| Helbig33 | 2009 | Germany | case report | 2 | VS | 0.00 | 49.5 (0.7) |
| Huang34 | 2021 | China | retrospective study | 10 | Brain or skull base radiation | 0.55 | 57.2 (6.2) |
| Huo35 | 2016 | China | retrospective case review | 2 | VS | 0.50 | 57 (4.2) |
| Irving36 | 1996 | U.K. | case report | 1 | SS | 0.00 | 33 (NA) |
| Jia37 | 2020 | France | retrospective case series | 5 | VS | 0.00 | 60.6 (12.2) |
| Longino38 | 2021 | U.S. | retrospective review | 6 | VS | 1.00 | 72.3 (14.8) |
| Low39 | 2006 | Singapore | retrospective case review | 4 | Brain or skull base radiation | 0.75 | 55.3 (6.9) |
| Lustig40 | 2006 | U.S. | retrospective case review | 2 | VS | 0.50 | 45.5 (6.4) |
| Medina41 | 2015 | Italy | case report | 1 | Brain or skull base radiation | 1.00 | 26 (NA) |
| Modest42 | 2015 | U.S. | case series | 5 | SS | 0.60 | 60.6 (12.6) |
| Mukherjee9 | 2013 | U.K. | retrospective review | 10 | VS | 0.20 | 56.2 (20.8) |
| Nogueira43 | 2012 | U.K. | case report | 1 | SS | 1.00 | 57 (NA) |
| North6 | 2016 | U.K. | retrospective case review | 6 | VS | 0.83 | 58.8 (15.0) |
| Omichi44 | 2016 | Japan | case report | 1 | SS | 1.00 | 38 (NA) |
| Pai45 | 2013 | U.K. | retrospective case series | 3 | VS | 0.00 | 68.9 (24.0) |
| Patel E.2 | 2021 | U.S. | retrospective review | 15 | VS | 0.60 | 57.3 (19.1) |
| Patel N.46 | 2021 | U.S. | retrospective chart review | 11 | VS | 0.00 | 48 (NA) |
| Pimentel47 | 2016 | Brazil | case report | 1 | VS | 1.00 | 50 (NA) |
| Pisa48 | 2017 | Canada | retrospective case review | 3 | VS | 0.33 | 51 (22.5) |
| Roehm49 | 2011 | U.S. | retrospective case review | 2 | VS | 0.00 | 56.5 (4.9) |
| Ryan50 | 2014 | Canada | case report | 1 | SS | 1.00 | 60 (NA) |
| Sugimoto51 | 2012 | Japan | case report | 1 | SS | 0.00 | 65 (NA) |
| Svrakic52 | 2017 | U.S. | case report | 2 | Neurosarcoidosis | 1.00 | 54 (NA) |
| Sydlowski53 | 2009 | U.S. | retrospective case report | 7 | SS | 0.00 | 57.9 (NA) |
| Tan54 | 2018 | China | retrospective case review | 4 | VS | 0.25 | 38.8 (14.7) |
| Tian55 | 2021 | Denmark | retrospective case series | 1 | VS | 1.00 | 54 (NA) |
| Tolisano56 | 2019 | U.S. | retrospective case series | 3 | VS | 0.00 | 26.3 (5.9) |
| Trotter57 | 2010 | Australia | retrospective case review | 3 | VS | 0.67 | 59 (22.3) |
| Urban58 | 2020 | U.S. | retrospective review | 6 | VS | 1.00 | 68.8 (16.0) |
| Wood59 | 2008 | New Zealand | retrospective case report | 2 | SS | 1.00 | 51.5 (2.1) |
| Yazama60 | 2021 | Japan | case report | 1 | SS | 1.00 | 30 (NA) |
| Yue61 | 2004 | China | retrospective chart review | 4 | Brain or skull base radiation | 0.75 | 56.8 (4.9) |

**Supplemental Table 2.** ROBINS-I assessment of bias.

|  |  |  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- | --- | --- |
| Author | Year | Confounding | Participant Selection | Classifiation of Interventions | Deviation From Intended Intervention | Missing Data | Outcome Measurements | Selection of Reported Result | Overall Bias |
| Alshehabi15 | 2019 | Critical | Low | Low | Moderate | Low | Low | Moderate | Critical |
| Amoodi16 | 2012 | Critical | Low | Low | Serious | Low | Serious | Moderate | Critical |
| Artukarslan17 | 2021 | Critical | Low | Low | Moderate | Low | Low | Moderate | Critical |
| Berrettini18 | 2012 | Critical | Low | Low | Serious | Low | Low | Moderate | Critical |
| Bird19 | 2010 | Critical | Serious | Low | Low | Low | Low | Moderate | Critical |
| Bittencourt20 | 2012 | Critical | Low | Low | Low | Low | Moderate | Moderate | Critical |
| Carlson21 Study **1** | 2012 | Critical | Low | Low | Moderate | Low | Serious | Moderate | Critical |
| Carlson22 Study **2** | 2016 | Critical | Low | Low | Low | Low | Moderate | Moderate | Critical |
| Clark75 | 2001 | Critical | Moderate | Low | Low | Low | Moderate | Moderate | Critical |
| Costello24 | 2016 | Critical | Moderate | Low | Low | Low | Moderate | Moderate | Critical |
| Deep5 | 2021 | Critical | Low | Low | Low | Low | Moderate | Moderate | Critical |
| Ebode25 | 2021 | Critical | Low | Low | Low | Low | Moderate | Moderate | Critical |
| Eitutis8 | 2021 | Critical | Low | Low | Low | Low | Low | Moderate | Critical |
| Formanek26 | 1998 | Critical | Moderate | Low | Low | Low | Moderate | Moderate | Critical |
| Fujimoto27 | 2007 | Critical | Moderate | Low | Low | Low | Moderate | Moderate | Critical |
| Greene28 | 2017 | Critical | Serious | Serious | Low | Low | Critical | Serious | Critical |
| Grover29 | 2011 | Critical | Moderate | Low | Low | Low | Moderate | Moderate | Critical |
| Halyur30 | 2021 | Critical | Moderate | Low | Low | Low | Moderate | Serious | Critical |
| Harris10 | 2017 | Critical | Low | Low | Low | Low | Moderate | Moderate | Critical |
| Hathaway31 | 2006 | Critical | Moderate | Low | Low | Low | Moderate | Moderate | Critical |
| Häußler32 | 2021 | Critical | Moderate | Low | Low | Low | Moderate | Moderate | Critical |
| Helbig33 | 2009 | Critical | Moderate | Low | Low | Low | Moderate | Moderate | Critical |
| Huang34 | 2021 | Critical | Low | Low | Low | Low | Low | Moderate | Critical |
| Huo35 | 2016 | Critical | Low | Low | Low | Low | Moderate | Moderate | Critical |
| Irving36 | 1996 | Critical | Moderate | Low | Low | Low | Moderate | Moderate | Critical |
| Jia37 | 2020 | Critical | Low | Low | Low | Low | Moderate | Moderate | Critical |
| Longino38 | 2021 | Critical | Low | Low | Low | Low | Moderate | Moderate | Critical |
| Low39 | 2006 | Critical | Low | Low | Low | Low | Moderate | Moderate | Critical |
| Lustig40 | 2006 | Critical | Low | Low | Low | Low | Moderate | Moderate | Critical |
| Medina41 | 2015 | Critical | Moderate | Low | Low | Low | Moderate | Moderate | Critical |
| Modest42 | 2015 | Critical | Low | Low | Low | Low | Moderate | Moderate | Critical |
| Mukherjee9 | 2013 | Critical | Low | Low | Low | Low | Low | Moderate | Critical |
| Nogueira43 | 2012 | Critical | Moderate | Low | Low | Low | Moderate | Moderate | Critical |
| North6 | 2016 | Critical | Low | Low | Low | Low | Moderate | Moderate | Critical |
| Omichi44 | 2016 | Critical | Moderate | Low | Low | Low | Moderate | Moderate | Critical |
| Pai45 | 2013 | Critical | Low | Low | Low | Low | Moderate | Moderate | Critical |
| Patel E.2 | 2021 | Critical | Low | Low | Low | Low | Moderate | Moderate | Critical |
| Patel N.46 | 2021 | Critical | Low | Low | Low | Low | Moderate | Moderate | Critical |
| Pimentel47 | 2016 | Critical | Moderate | Low | Low | Low | Moderate | Moderate | Critical |
| Pisa48 | 2017 | Critical | Low | Low | Low | Low | Low | Moderate | Critical |
| Roehm49 | 2011 | Critical | Low | Low | Low | Low | Moderate | Moderate | Critical |
| Ryan50 | 2014 | Critical | Moderate | Low | Low | Low | Moderate | Moderate | Critical |
| Sugimoto51 | 2012 | Critical | Moderate | Low | Low | Low | Moderate | Moderate | Critical |
| Svrakic52 | 2017 | Critical | Moderate | Low | Low | Low | Moderate | Moderate | Critical |
| Sydlowski53 | 2009 | Critical | Low | Low | Low | Low | Moderate | Moderate | Critical |
| Tan54 | 2018 | Critical | Low | Low | Low | Low | Moderate | Moderate | Critical |
| Tian55 | 2021 | Critical | Moderate | Low | Low | Low | Moderate | Moderate | Critical |
| Tolisano56 | 2019 | Critical | Low | Low | Low | Low | Moderate | Moderate | Critical |
| Trotter57 | 2010 | Critical | Low | Low | Low | Low | Serious | Serious | Critical |
| Urban58 | 2020 | Critical | Low | Low | Low | Low | Moderate | Moderate | Critical |
| Wood59 | 2008 | Critical | Moderate | Low | Low | Low | Serious | Serious | Critical |
| Yazama60 | 2021 | Critical | Moderate | Low | Low | Low | Serious | Serious | Critical |
| Yue61 | 2004 | Critical | Low | Low | Low | Low | Low | Moderate | Critical |

**Supplemental Table 3.** Description of patients who did not achieve immediate or sustained benefit from cochlear implantation.

|  |  |  |  |
| --- | --- | --- | --- |
| Author | Pathology | Number of patients without immediate or sustained benefit | Details |
| Alshehabi15 | SS | 3 | These three patients had no improvement in speech perception from their cochlear implants, although no additional details were provided. |
| Artukarslan17 | SS | 2 | 68-year-old female with superficial siderosis had no speech understanding after implantation, despite evidence of normal device integrity. Authors suspect poor performance was related to extent and location of hemosiderin deposits. A second patient, a 59-year-old male, received a CI and had initial speech scores of 70%, which dropped over a 9-year period to 30%, progression was also linked to progressive disease. |
| Carlson21 | VS | 1 | 44-year-old blind female with NF2, who underwent SRS (marginal and maximal doses of 20 and 40 Gy respectively) 20 years prior to cochlear implantation. The patient reported immediate and substantial benefit after CI, however, one year after surgery, she lost all sound perception capability. Authors list potential causes including delayed radiation injury, unidentified device malfunction, or increasing tumor burden, but do not provide any further details. |
| Deep5 | VS | 1 | 21-year-old female with NF2, who received a CI on the side of an observed, 3 cm VS and reported minimal subjective benefit with only limited access to environmental sounds. Authors suspect possible larger tumor size as a potential reason for poor outcomes but cannot draw conclusions due to small patient numbers. |
| Eitutis8 | VS | 1 | 41-year-old with a 16 mm previously irradiated VS received no benefit from CI, which was performed 16 years after radiation therapy. Possible reason for poor outcomes was presence of tip foldover. |
| Grover29 | SS | 1 | 54-year-old male with superficial siderosis, dementia, seizures, visual impairment, and no word recognition to speech reading prior to implantation required very high current levels for stimulation and had no auditory perception after CI. |
| Huo35 | VS | 1 | 60-year-old female with a 9 mm untreated sporadic vestibular schwannoma who post-operatively could only detect environmental sounds and had improved lip reading, but her CI worsened her tinnitus and thus was not a regular user. No clear cause for poor performance was identified. |
| Modest42 | SS | 2 | Two patients with superficial siderosis did not demonstrate sustained benefit due to disease progression 18 and 24 months after implantation. |
| Mukherjee9 | VS | 1 | 20-year-old female with advanced NF2, and known for several cranial nerve palsies and a left-sided 3.7 cm tumor which underwent GKRS 4 years prior to implantation, did not achieve any speech discrimination, but was able to hear her dog bark which she ultimately reported as a significant improvement in her quality of life. |
| Patel2 | Brain or skull base radiation | 2 | 65-year-old male with sporadic vestibular schwannoma did not show benefit in speech discrimination and was a non-user of his device, but no additional details were provided.  20-year-old male with disseminated CNS germinoma who underwent chemotherapy (carboplatin, etoposide) and proton-beam radiation prior to implantation. Promontory stimulation showed no response pre-operatively, but the implant was performed nonetheless, patient did not achieve any sound perception. |
| Tan54 | VS | 1 | 44-year-old female with NF2 with a 3.1 cm untreated tumor who did have a slight improvement in speech perception but subjectively was only able to identify voice modulation and felt the device interfering with her contralateral hearing aid and opted to keep the device as a “sleeper”. |
| Urban58 | VS | 1 | 38-year-old male with NF2 and a 28 mm irradiated VS had immediate benefit from his cochlear implant but suffered from delayed hearing loss 18 months post-operatively and discontinued use of his device. No further details are provided relating to performance deterioration. |
| Wood59 | SS | 2 | 53-year-old male demonstrated improved speech discrimination post-operatively but gradually had declined performance. Progressive neural deterioration was suspected as the etiology of the hearing loss and at one-year post-op he could detect environmental sounds only.  50-year-old male who at first demonstrated improvement in speech perception (HINT AV 77.3% and AA 19%, CNC AV 60% and AA 0%), but one month after processor fitting, demonstrated substantial deterioration in global functioning and balance and was determined to have dementia from his superficial siderosis leading to decline in performance. |
| Yazama60 | SS | 1 | 30-year-old male with superficial siderosis and history of anaplastic astrocytoma of the midbrain showed no improvement in speech perception after CI. Authors attributed the poor results due to poor usage and poor rehabilitation compliance. |