CHAPTER TEN

EARS

The first time you see the inside of the ear is a letdown: the real machinery is hidden deep inside the skull, encased in a wall of bone. Once you open the skull and remove the brain, you need to chip with a chisel to remove that wall. If you are really good, or very lucky, you'll make the right stroke and see it—the inner ear. It resembles the kind of tiny coiled snail shell you find in the dirt in your lawn.

The ear may not look like much, but it is a wonderful Rube Goldberg contraption. When we hear, sound waves are funneled into the outside flap, the external ear. The sound waves enter the ear and make the eardrum rattle. The eardrum is attached to three little bones, which shake along with it. One of these ear bones is attached to the snail-shell structure by a kind of plunger. The shaking of the ear bone causes the plunger to go up and down. This causes some gel inside the snail shell to move around. Swishing gel bends nerves, which send a signal to the brain, which interprets it as sound. Next time you are at a concert, just imagine all the stuff flying around in your head. This structure allows us to distinguish three parts to the ear: external, middle, and inner. The external ear is the visible part. The middle contains the little ear bones. Finally, the inner ear consists of the nerves, the gel, and the tissues that surround them. These three components of ears enable us to structure our discussion in a very convenient way.



Of the three parts of our ear—the outer, middle, and inner—the inner ear is the most ancient and the part that controls the nerve impulses sent to the brain.

The part of the ear we can see, the flap on which we hang our glasses, is a relatively new evolutionary addition to bodies. Confirm this on your next trip to the aquarium or zoo. How many sharks, bony fish, amphibians, and reptiles have external ears? The pinna—the flap of the external ear —is found only in mammals. Some amphibians and reptiles have visible external ears, but they have no pinna. Often the external ear is only a membrane that looks like the top of a drum.

The elegance of our connection to sharks and bony fish is revealed when we look inside our ears. Ears might seem an unlikely place for a human-shark connection, especially since sharks don't have ears. But the connection is there. Let's start with the ear bones.

THE MIDDLE EAR—THE THREE EAR BONES

Mammals are very special. With hair and milk-producing glands, we can easily be distinguished from other creatures. It surprises most people to learn that some of the most distinctive traits of mammals lie inside the ear. The bones of the mammalian middle ear are like those of no other animal: mammals have three bones, whereas reptiles and amphibians have only one. Fish have none at all. Where did our middle ear bones come from?

Some anatomy: recall that our three middle ear bones are known as the malleus, incus, and stapes. As we've seen, each of these ear bones is derived from the gill arches: the stapes from the first arch, and the malleus and incus from the second arch. It is here that our story begins.

In 1837, the German anatomist Karl Reichert was looking at embryos of mammals and reptiles to understand how the skull forms. He followed the gill arches of different species to understand where they ended up in the various skulls. As he did this again and again, he found something that appeared not to make any sense: two of the ear bones in the mammals corresponded to pieces of the jaw in the reptiles. Reichert could not believe his eyes, and his monograph reveals his excitement. As he describes the ear-jaw comparison, his prose departs from the normally staid description of nineteenth-century anatomy to express shock, even wonderment, at this discovery. The conclusion was inescapable: the same gill arch that formed part of the jaw of a reptile formed ear bones in mammals. Reichert proposed a notion that even he could barely believe-that parts of the ears of mammals are the same thing as the jaws of reptiles. Things get more difficult when we realize that Reichert proposed this several decades before Darwin propounded his notion of a family tree for life. What does it mean to call structures in two different species "the same" without a notion of evolution?

Much later, in 1910 and 1912, the German anatomist Ernst Gaupp picked up on Reichert's work and published an exhaustive study on the embryology of mammalian ears. Gaupp provided more detail and, given the times, interpreted Reichert's work in an evolutionary framework. Gaupp's story went like this: the three middle ear bones reveal the tie between reptiles and mammals. The single bone in the reptilian middle ear is the same as the stapes of mammals; both are second-arch derivatives. The explosive bit of information, though, was that the two other middle ear bones of mammals—the malleus and the incusevolved from bones set in the back of the reptilian jaw. If this was indeed the case, then the fossil record should show bones shifting from the jaw to the ear during the origin of mammals. The problem was that Gaupp worked only on living creatures and didn't fully appreciate the role that fossils could play in his theory.

Beginning in the 1840s a number of new kinds of fossil creatures were becoming known from discoveries in South Africa and Russia. Often abundantly preserved, whole skeletons of dog-size animals were being unearthed. As they were discovered, many of them were crated and shipped to Richard Owen in London for identification and analysis. Owen was struck that these creatures had a mélange of features. Parts of their skeleton looked reptilelike. Other parts, notably their teeth, looked like mammals. And these were not isolated finds. It turns out that these "mammal-like reptiles" were the most common skeletons being uncovered at many fossil sites. Not only were they very common, there were many kinds. In the years after Owen, these mammal-like reptiles became known from other parts of the world and from several different time periods in earth history. They formed a beautiful transitional series in the fossil record between reptile and mammal.

Until 1913, embryologists and paleontologists were working in isolation from one another. At this time, the American paleontologist W. K. Gregory, of the American Museum of Natural History, saw an important link between Gaupp's embryos and the African fossils. The most reptilian of the mammal-like reptiles had only a single bone in its middle ear; like other reptiles, it had a jaw composed of many bones. Something remarkable was revealed as Gregory looked at the successively more mammalian mammal-like reptiles, something that would have floored Reichert had he been alive: a continuum of forms showing beyond doubt that over time the bones at the back of the reptilian jaw got smaller and smaller, until they ultimately lay in the middle ear of mammals. The malleus and incus did indeed evolve from jawbones. What Reichert and Gaupp observed in embryos was buried in the fossil record all along, just waiting to be discovered.

Why would mammals need a three-boned middle ear? This little linkage forms a lever system that allows mammals to hear higher-frequency sounds than animals with a single middle ear bone. The origin of mammals involved not only new patterns of chewing, as we saw in Chapter 4, but new ways of hearing. In fact, this shift was accomplished not by evolving new bones per se, but by repurposing existing ones. Bones originally used by reptiles to chew evolved in mammals to assist in hearing.

So much for the malleus and incus. Where, though, does the stapes come from?

If I simply showed you an adult human and a shark, you would never guess that this tiny bone deep inside a human's ear is the same thing as a large rod in the upper jaw of a fish. Yet, developmentally, these bones are the same thing. The stapes is a second-arch bone, as is the corresponding bone in a shark and a fish—the hyomandibula. But the hyomandibula is not an ear bone; recall that fish and sharks do not have ears. In our aquatic cousins, this bone is a large rod that connects the upper jaw to the braincase. Despite the apparent differences in the function and shape of these bones, the similarities between the hyomandibula and the stapes extend even to the nerves that supply them. The key nerve for the functioning of both bones is the second-arch nerve, the facial nerve. We thus have a situation where two very different bones have similar developmental origins and patterns of innervation. Is there an explanation for this?

Again, we look to the fossils. As we trace the hyomandibula from sharks to creatures like *Tiktaalik* to amphibians, we can see how it gets smaller and smaller, ultimately shifting position from the upper jaw to play a role in hearing. The name changes, too. When it is big and supporting the jaw, we call it a hyomandibula; when it is small and functions in hearing, it is known as a stapes. This shift happened when the descendants of fish began to walk on land. Hearing in water is different from hearing on land, and the small size and position of the stapes makes it ideal for picking up vibrations in air. The new ability came about by modifying the upper jawbone of a fish.

We can trace bones from gill arches to our ears, first during the transition from fish to amphibian (right), and later during the shift from reptile to mammal (left).

Our middle ear contains a record of two of the great transformations in the history of life. The origin of our stapes, and its transformation from a jaw support bone to an ear bone, began when fish started to walk on land. The other big event took place during the origin of mammals, when bones at the back of a reptile jaw became our malleus and incus.

Now let's go further inside the ear—to the inner ear.

THE INNER EAR—GELS MOVING AND HAIRS BENDING

Move through the external ear, go deeper inside, past the eardrum and three middle ear bones, and you end up deep inside the skull. Here you will find the inner ear—tubes and some gel-filled sacs. In humans, as in other mammals, the bony tubes take the snail-shell shape that is so strikingly apparent in the anatomy lab.

The inner ear has different parts dedicated to different functions. One part is used in hearing, another in telling us which way our head is tilted, and still another in recording how fast our head is accelerating or stopping. In carrying out each of these functions, the inner ear works in roughly the same way.

The several parts of the inner ear are filled with a gel that can move. Specialized nerve cells send hairlike projections into this gel. When the gel moves, the hairs on the ends of the nerve cells bend. When these hairs bend, the nerve cells send an electrical impulse to the brain, where it is recorded as sound, position, or acceleration.

Each time you tilt your head, the tiny rocks on the fluid-filled sacs move. In doing so, they bend nerve endings inside the sacs and cause an impulse to be sent to your brain saying "Your head is tilted."

To envision the structure that tells us where our head is in space, imagine a Statue of Liberty snow globe. The snow globe is made of plastic and filled with gel. When you shake it, the gel moves and the "snow" falls on Lady Liberty. Now imagine a snow globe made of a flexible membrane. Pick it up and tilt it, and the whole thing will flop about, causing the gel inside to swish around. This, on a much smaller scale, is what we have inside our ears. When we bend our heads, these contraptions flop around, causing the usual chain of events: the gel inside swishes, the hair projections on the nerves bend, and an impulse is sent back to our brains.

In us, this whole system is made even more sensitive by the presence of tiny rock-like structures on top of the membrane. As we bend our heads, the rocks accentuate the flopping of the membrane, causing the gel to move even more. This increases the sensitivity of the system, enabling us to perceive small differences in position. Tilt your head, and little rocks inside your skull move.

You can probably imagine how tough it would be to live in outer space. Our sensors are tuned to work in the earth's gravity, not in a gravity-free space capsule. Floating around, our eyes recording one version of up and down, our inner ear sensors totally confused, it is all too easy to get sick. Space sickness has been a real problem for these very reasons.

The way we perceive acceleration is based on yet another part of our inner ear, connected to the previous two. There are three gel-filled tubes inside the ear; each time we accelerate or stop, the gel inside the tubes moves, causing the nerve cells to bend and stimulate a current.

The whole system we use to perceive position and acceleration is connected to our eye muscles. The motion of our eyes is controlled by eight small muscles attached to

the side walls of the eyeball. The muscles contract to move the eye up, down, left, and right. We can move our eyes voluntarily by contracting these muscles each time we decide to look in a new direction; but some of the most fascinating properties of these muscles relate to their involuntary action. They move our eyes all the time, without our even thinking about it.

To appreciate the sensitivity of this eye-muscle link, move your head back and forth while looking at the page. Keep your eyes fixed in one place as you move your head.

What happened during this experiment? Your eyes stayed fixed on a single point while your head moved. This motion is so commonplace that we take it for granted, but it is incredibly complex. Each of the eight muscles in both eyes is responding to the movement of the head. Sensors in your head, which I'll describe in the next section, record the direction and velocity of your head's movement. These signals are carried to the brain, which then sends out signals telling your eye muscles to fire. Think about that the next time you fix your gaze as your head is moving. This system can misfire, and misfires have much to tell us about our general well-being.

Every time we accelerate, fluid in the inner ear swishes. The swish is transformed into a nerve impulse that is sent to the brain.

An easy way to understand the inner ear–eye connection is to interfere with it. One way humans do this is to imbibe too much alcohol. Drinking too much ethanol leads us to do silly things because our inhibitions are lowered. Drinking *way* too much gives us the spins. And the spins often predict a lousy morning ahead, hungover, with more spins, nausea, and headache.

When we drink too much, we are putting lots of ethanol into our bloodstream, but the fluid inside our ear tubes initially contains very little. As time passes, however, the alcohol diffuses from our blood into the gel of the inner ear. Alcohol is lighter than the gel, so the result of the diffusion is like the result of pouring alcohol into a glass of olive oil. Just as the oil moves around in the glass as the alcohol enters, so the gel inside our ear swirls. The convection wreaks havoc on the intemperate among us. Our hair cells are stimulated and our brain thinks we are moving. But we are not moving; we are slumped in a corner or hunched on a barstool. Our brain has been tricked.

The problem extends to our eyes. Our brain thinks we are spinning, and it passes this information to our eye muscles. The eyes twitch in one direction (usually to the right) as we try to track an object moving from side to side. If you prop open the eyes of someone who is stone drunk, you might see this stereotypical twitch, called nystagmus. Police know this well, and often look for nystagmus in people whom they have stopped for driving erratically.

Massive hangovers involve a slightly different response. The day after the binge, your liver has done a remarkably efficient job of removing the alcohol from your bloodstream. Too efficient, for we still have alcohol in the tubes in our ears. That alcohol then diffuses from the gel back into the bloodstream, and in doing so it once more sets the gel in motion: the spins again. Take the same heavy drinker whose eyes you saw twitch to the right the night before and look at him during the hangover. His eyes might still twitch, but in the opposite direction.

We can thank our shared history with sharks and fish for this. If you have ever tried to catch a trout, then you have come up against an organ that is likely an antecedent to our inner ear. As every fisherman knows, trout hold only in certain parts of a stream, typically spots where they can get the best meal while avoiding predators. Often such places are in the shade and in the eddies of the stream's current. Great places for big fish to hold are behind big rocks or fallen logs. Trout, like all fish, have a mechanism that allows them to sense the current and the motion of the water around them, almost like a sense of touch.

Within the skin and bones of the fish, arranged in lines that run the length of the body and head, are small organs with sensory receptors. These receptors lie in small bundles from which they send small hair-like projections into a jelly-filled sac called a neuromast organ. It helps to think of the snow globe Statue of Liberty again. A neuromast organ is like a tiny one of these, with nerves projecting inside. When the water flows around the fish, it deforms this small sac, thereby bending the hair-like projections of the nerve. Much like the whole system in our ears, this apparatus then sends a signal back to the brain and gives the fish a sense of what the water is doing around them. Sharks and fish can discern the direction in which the water is flowing, and some sharks can even detect distortions of the water, such as are produced by other fish swimming near them. We used a version of this system when we moved our head with a fixed gaze, and we saw it go awry when we propped open the eyes of the inebriated individual at the start of this section. If the ancestor we have in common with sharks and fish had used some other kind of inner ear gel, say one that does not swirl when alcohol is added, we would never spin when drunk.

If you think of our inner ears and neuromast organs as versions of the same thing, you would not be far off. Both come from the same sort of tissue during development, and they share a similar structure. But which came first: neuromasts or inner ears? Here the evidence gets sketchy. If you look at some of the earliest fossils with heads, creatures about 500 million years old, you'll find little pits in their external armor that suggest they had neuromast organs. Unfortunately, we do not know much about the inner ears of these creatures because the preservation of that area of the head is wanting. Until more evidence rolls in, we are left with one of two alternatives: either our inner ears arose from neuromast organs or the other way around. Both scenarios, at their core, reflect a principle we've seen at work in other parts of the body. Organs can come about for one function, only to be repurposed over time for any number of new uses.

In our own ears, there occurred an expansion of the inner ear. The part of our inner ear devoted to hearing is, as in other mammals, huge and coiled. More primitive creatures, such as amphibians and reptiles, have a simple uncoiled inner ear. Clearly, our mammalian forebears obtained a new and better type of hearing. The same is true for the structures that perceive acceleration. We have three canals to record acceleration because we perceive space in three dimensions. The earliest known fish with these canals, a kind of jawless fish like a hagfish, has only one. Then, in other primitive fish, we see two. Finally, most modern fish, and other vertebrates, have three, like us.

We have seen that our inner ear has a history that can be traced to the earliest fish. Remarkably, the neurons inside the gel of our ears have an even more ancient history.

These neurons, called hair cells, have special features that are seen in no other neuron. With fine hair-like projections, consisting of one long "hair" and a series of smaller ones, these neurons lie with a fixed orientation in our inner ear and in a fish's neuromast organ. Recently, people have searched for these cells in other creatures, and have found them not only in animals that do not have sense organs like ours at all but also in animals that have no heads. They are seen in creatures like *Amphioxus*, which we met in Chapter 5, that have no ears, eyes, heads, or skulls. Hair cells, then, were around doing other things before our sense organs even hit the scene.

A primitive version of part of our inner ear is embedded in the skin of fish. Small sacs—the neuromasts—are distributed around the body. When they bend, they give the fish information about how the flow of water is changing.

All this is recorded in our genes, of course. If humans or mice have a mutation that knocks out a gene called *Pax 2*, the inner ear fails to form properly. *Pax 2* is active in the ear region and appears to start a chain reaction of gene activity that leads to the development of the inner ear. Go fishing for this gene in more primitive animals and we find *Pax 2* active in the head and, lo and behold, in the neuromasts.

The spinning drunk and the fish's water-sensing organs have common genes: evidence of a common history.

JELLYFISH AND THE ORIGINS OF EYES AND EARS

Just like *Pax 6*, which we discussed earlier in connection with eyes, *Pax 2* in ears is a major gene, essential for proper development. Interestingly, a link between *Pax 2* and *Pax 6* suggests that ears and eyes might have had a very ancient common history.

This is where the box jellyfish enters our story. Well known to swimmers in Australia because they have particularly poisonous venom, these jellyfish are different from most others in that they have eyes, more than twenty of them. Most of these eyes are simple pits spread over the jellyfish's epidermis. Other eyes on the body are strikingly similar to our own, with a kind of cornea, a lens, even a nervous structure like ours.

Jellyfish do not have either *Pax 6* or *Pax 2:* they arose before those genes hit the scene. But in the box jellyfish's genes we see something remarkable. The gene that forms the eyes is not *Pax 6,* as we'd expect, but a sort of mosaic that has the structure of *both Pax 6* and *Pax 2.* In other words, this gene looks like a primitive version of other animals' *Pax 6* and *Pax 2.*

The major genes that control our eye and ear correspond to a single gene in more primitive creatures, such as jellyfish. You're probably thinking, So what? The ancient connection between ear and eye genes helps to make sense of things we see in hospital clinics today: a number of human birth defects affect *both* the eyes and the inner ear. All this is a reflection of our deep connections to primitive creatures like the stinging box jellyfish.