



Charles V (1500-1558) Charles I of Spain  
1519-1556, Holy Roman Empire 1519-1558.  
Portrait engraving showing his prominent lower  
'Habsburg' deformed jaw.  
Credit: UniversallimagesGroup / Contributor

# Small, medium, and large:

## Growth abnormalities of the mandible

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“**T**he Remarkable Life of Erwin Schrödinger’s *What Is Life?*” in the Autumn 2015 issue of *The Pharos* uses, “the infamous Habsburg lip, faithfully preserved throughout the long dynasty,” as an example of genetic fidelity.

This genetic fidelity—mandibular hyperplasia, known as prognathism or osseous Class III malocclusion—is commonly referred to as the Habsburg jaw. The Habsburg’s lower lips were large, as were their tongues, but their lower jaws (mandible) were exceptionally hypertrophied, jutting out from under their upper jaws (maxilla).

Many human diseases, deformities and disabilities come

in three sizes—small, medium and large. Disharmony of size or shape between the upper and lower jaws in any dimension—height, width, or length—can create serious functional problems that interfere with mastication, and cause esthetic issues.

The ideal small may be only a few millimeters of discrepancy between the jaws and the intermeshing of the teeth, basically an issue obvious only to an orthodontist. Medium differences between the jaws may be more cosmetically obvious and may compromise an individual’s ability to functionally masticate food. Large denotes asymmetry or disharmony, and seriously affects speech and swallowing.

For the ancient Greeks, beauty was synonymous with symmetry—Plato wrote of golden proportions—but very few humans have ever met the ideal of perfect facial harmony.<sup>1</sup>



Portrait of Charles V, Holy Roman Emperor, by Jan Cornelisz Vermeyen (manner of), c. 1530. Courtesy of the Rijksmuseum



Philip IV (1605-1665) king of Spain, 1628. His long, pronounced Hapsburg jaw is evident in this portrait by Diego Velasquez (1599-1660), Prado, Madrid.

Credit: UniversallImagesGroup/Contributor

Perfect is in the eye of the beholder, Helen in the eyes of Paris, or golden Aphrodite.

The 16th century Hapsburg dynasty of Spain, Austria, and the Holy Roman Empire suffered a familial disorder of the large variety—lower jaws protruding far in front of smaller upper jaws. This deformity is apparent in paintings of generations of Hapsburgs, especially the Titian portraits of Philip II of Spain, and Charles V, Holy Roman Emperor.

The Hapsburgs' jaws may or may not have been a social issue, having great impact on the history of the Spanish Empire. However, it is said that contemporaries found Philip II "difficult to understand...often unintelligible."<sup>2</sup> And, it is documented that Charles V's "Hapsburg blood was apparent in his hanging jaw and gaping look."<sup>2</sup>

### The anguish of the disfigurement

Disfigurement, be it genetic, traumatic, or caused by ablative cancer surgery, can create heavy personal and social burdens. Lucy Grealy, the author and poet who suffered a childhood jaw tumor described her anguish:

My pleasure at the sight of the children didn't last long, however. I knew what was coming...they'd notice me. Half my jaw missing, which gave my face a strange triangular shape, accentuated by the fact that I was unable to keep my mouth completely closed.<sup>3</sup>

Grealy's emotional suffering was devastating and incomprehensible. Her friend, Ann Patchett tells the story

of Grealy's decline into depression, alcohol, drugs and death in the book "Truth and Beauty:"

We could see her face clearly. It was always changing, swollen after a surgery or sinking in on itself after a surgery had failed. One year she walked with a cane and someone told me it was because they had taken a chunk of her hip to grind up a graft into her jaw.<sup>4</sup>

In the best of all worlds, talent, education, and health should lead to social and economic success, and a reasonable measure of happiness. However, numerous studies have revealed that facial appearance influences self-image as well as other's perceptions, thus affecting nearly every aspect of life, from job promotion to finding a mate.<sup>5,6</sup>

### Surgical advancements

The first successful surgical attempt to correct growth abnormalities of the mandible was by Thomas Houlihan, a mid-19th century American surgeon who removed bone segments bilaterally, and wired the patient's upper and lower teeth together to stabilize the healing bone. The surgery was done with crude anesthesia, and no antibiotics.

A series of innovative surgeons improved on Houlihan's procedure throughout the decades, until the 1960s when Professor Hugo Obwegeser of Zurich perfected techniques that could alter the size, shape, and position of both jaws simultaneously without bone removal or surgical scars on the face.

Later, the use of small orthopedic titanium plates and screws obviated the need to wire the mouth shut postoperatively, a major technological leap in the evolution of successful reconstructive jaw surgery.

### The regeneration of Billy

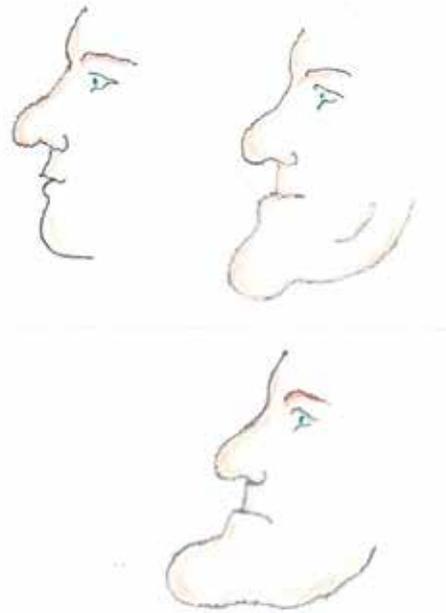
"Billy" was 19 years old, a high school dropout, and a recidivistic car thief incarcerated by the Connecticut criminal justice system. He came from a dysfunctional family described by the prison psychologist as uncaring and unwitting. His mandible was huge, even larger than a Hapsburg's. He had lost all of his teeth due to neglect and poverty.

Billy's jaw was so large that functional dentures could not be fabricated. His hyperplastic-prognathic jaw was not that of pituitary gigantism, non-pituitary gigantism (Soto's syndrome), or any of the numerous other syndromic conditions associated with jaw abnormalities, and he was not a Hapsburg.

Years of social ostracism and cruel jokes had produced an angry, isolated, sullen, introverted young man who was so taciturn that he bordered on mute. The psychologist suggested that "fixing" Billy's face might create some self-esteem, and perhaps open new vistas for him. Billy expressed interest in being able to "chew some meat and maybe not have the girls turn away."

I realized that Billy's anger and rebellion may be just a *cri de cœur* that he couldn't articulate.

The surgery went well, and form and function were restored. Early in his post-operative period, Billy's personality completely changed, and he became energized, almost loquacious, interacting with and pursuing every young nurse who walked by his room. He often left his room to read magazines in the hospital library. I was asked if a guard should be placed outside his room, or if he should be placed in shackles, but he wasn't going to escape, not this new Billy. He no longer was displaying rebellious behavior problems.



When he returned to the penitentiary the prison dentist created dentures for him, allowing Billy to have standard meals rather than pureed prison fare.

During subsequent visits to the hospital clinic Billy was cooperative, in good humor, and eager to finish his sentence in the penitentiary. He was considering completing high school or applying to an auto mechanic training program, and "maybe buy a car!" he joked.

What became of Billy? Perhaps on a straight path on his journey through life, a steady job, the serene joy of love and a family, the satisfaction of food? I'll never know. I can only hope. But I know for sure, he was healthier, had improved self-esteem, and was rid of his large jaw.

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