

## Müllerian Agenesis and Beyond: A Comprehensive Analysis of Rokitansky-Küster-Hauser Syndrome

Rebeca Álvarez Fernández<sup>1</sup>, Abril Mariana Abraham Mexicano<sup>2</sup>, Emmanuel Bautista Perea<sup>3</sup>, Blanca Areni González Solís<sup>4</sup>, Imanol Guadalupe Martínez<sup>5</sup>, Luis Enrique Morales Montoya<sup>6</sup>

<sup>1</sup>Universidad Autónoma de Guadalajara. Guadalajara, Jalisco, México.

<sup>2</sup>Universidad de Guanajuato. Guanajuato, México.

<sup>3</sup>Universidad Nacional Autónoma de México. Facultad de estudios Superiores Iztacala.

<sup>4</sup>Universidad del Valle de México. Campus Reynosa.

<sup>5</sup>Instituto Politécnico Nacional, Escuela Superior de Medicina.

<sup>6</sup>Universidad Autónoma de Sinaloa, Culiacán, Sinaloa. México

### ABSTRACT

Rokitansky-Küster-Hauser syndrome (RKH), also known as Müllerian agenesis, represents a rare congenital disorder characterized by the absence or underdevelopment of the müllerian duct structures in phenotypic females. This condition manifests as primary amenorrhea, with a typical constellation of associated anomalies encompassing the urogenital and skeletal systems. Despite its rarity, RKH poses significant physical, psychological, and reproductive challenges to affected individuals. This comprehensive article endeavors to elucidate the complex clinical spectrum, pathogenesis, diagnostic modalities, and multidisciplinary management strategies for Rokitansky-Küster-Hauser syndrome.

**KEYWORDS:** syndrome, Rokitansky-Küster-Hauser, Mullerian, agenesis

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### INTRODUCTION

Rokitansky-Küster-Hauser syndrome (RKH), an eponymous term derived from the names of its discoverers, represents a notable enigma within the realm of congenital reproductive anomalies. This condition is characterized by the absence or rudimentary development of the müllerian duct structures, which culminates in the absence of a functional uterus and upper two-thirds of the vagina. The classic clinical presentation of primary amenorrhea and normal secondary sexual characteristics in phenotypic females conceals a plethora of intricate challenges.<sup>1,2</sup>

The etiopathogenesis of RKH remains a topic of active exploration, involving a delicate interplay of genetic, hormonal, and embryological factors. Beyond its reproductive implications, RKH often accompanies a spectrum of urogenital, skeletal, and occasionally renal anomalies, necessitating a multidisciplinary approach to clinical management.<sup>1,2</sup>

This article aims to provide a comprehensive understanding of Rokitansky-Küster-Hauser syndrome by delving into its clinical presentation, diagnostic methodologies, genetic

underpinnings, and the intricacies of its management. It is our aspiration that this exploration will contribute to the dissemination of knowledge and further advance the quality of care provided to individuals grappling with this rare and complex syndrome.<sup>1,2</sup>

### EPIDEMIOLOGY

Rokitansky-Küster-Hauser Syndrome (RKH), an exceedingly rare and complex congenital anomaly, presents a distinct epidemiological profile, characterized by its sporadic occurrence and considerable phenotypic heterogeneity. Understanding the epidemiology of RKH is fundamental to both healthcare planning and the optimization of clinical care for individuals affected by this syndrome. This article elucidates the multifaceted epidemiological aspects of RKH, delving into its incidence, prevalence, risk factors, geographical distribution, and potential genetic underpinnings.<sup>2,3</sup>

Incidence and Prevalence:

RKH, also known as Müllerian agenesis, is a condition predominantly encountered in phenotypic females, and its

**Corresponding Author: Rebeca Álvarez Fernández**

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precise incidence has proven elusive to ascertain accurately due to its rarity. The estimated incidence of RKH ranges from 1 in 4,000 to 1 in 10,000 female births, although these figures can vary across different populations and might be underreported due to misdiagnosis or underdiagnosis. The prevalence of RKH, defined as the total number of affected individuals within a specific population at a given point in time, is influenced by factors such as genetic predisposition and ethnic background.<sup>2,3</sup>

### Risk Factors:

The etiological factors contributing to RKH remain elusive, with no single causative agent identified to date. Nonetheless, there is evidence to suggest a potential genetic component, as familial clustering of cases has been reported, and studies exploring a genetic basis through linkage analyses and candidate gene studies are ongoing. Additionally, associations with maternal exposures during pregnancy and the use of certain medications have been hypothesized as potential risk factors, albeit inconclusively.<sup>2,3</sup>

### Geographical Distribution:

RKH does not exhibit a significant geographical predilection, and cases have been reported across diverse populations and regions. However, the availability of healthcare resources and diagnostic expertise may influence the detection and reporting of RKH, thereby affecting its apparent distribution. It is crucial to acknowledge that the syndrome may be underrepresented in regions with limited access to specialized healthcare services and expertise in rare congenital anomalies.<sup>2,3</sup>

### Genetic Underpinnings:

Emerging evidence supports the notion of genetic underpinnings in the pathogenesis of RKH. Mutations in several genes, such as WNT4 and HOXA genes, have been implicated in Müllerian duct development and may play a role in the etiology of RKH. These genetic insights could open new avenues for early diagnosis, genetic counseling, and potential therapeutic interventions in the future.<sup>2,3</sup>

While Rokitansky-Küster-Hauser Syndrome remains a rare and enigmatic condition, its epidemiological landscape is gradually coming into focus. Ongoing research endeavors are shedding light on the potential genetic factors underlying its pathogenesis, and improved diagnostic techniques are enhancing case detection. Comprehensive epidemiological studies are essential not only for understanding the prevalence and risk factors associated with RKH but also for the development of targeted healthcare interventions that can better support individuals living with this complex congenital anomaly.<sup>2,3</sup>

## RELEVANCE

Rokitansky-Küster-Hauser Syndrome (RKH), also known as Müllerian agenesis, stands as a condition of remarkable clinical and scientific relevance in the field of medicine. This congenital disorder, characterized by the underdevelopment

or complete absence of the müllerian duct structures, has gained substantial recognition and importance due to several pivotal facets that warrant in-depth exploration.<sup>3,4</sup>

### 1. Clinical Significance:

RKH's foremost relevance lies in its profound impact on the lives of affected individuals. The hallmark of primary amenorrhea in phenotypic females brings about intricate challenges, both psychological and physiological. The absence of a functional uterus and upper vaginal tract poses profound concerns related to fertility, sexual function, and psychosocial well-being. As such, understanding and addressing these clinical ramifications is of paramount importance.<sup>3,4</sup>

### 2. Diagnostic Challenges and Advances:

The diagnosis of RKH can be intricate, necessitating a multidisciplinary approach encompassing gynecologists, endocrinologists, radiologists, and genetic counselors. The advent of advanced diagnostic modalities, such as magnetic resonance imaging (MRI) and genetic testing, has substantially improved the accuracy and timeliness of diagnosis, underscoring the relevance of staying abreast of evolving diagnostic tools.<sup>3,4</sup>

### 3. Genetic Insights:

RKH has increasingly become the focus of genetic research. The identification of candidate genes associated with müllerian duct development, such as WNT4 and HOXA, has opened up new avenues for understanding its genetic underpinnings. This genetic insight is not only crucial for elucidating the syndrome's etiology but also for advancing the potential for genetic counseling and targeted interventions.<sup>3,4</sup>

### 4. Reproductive and Fertility Concerns:

RKH significantly impacts reproductive health, as affected individuals often desire to conceive. The relevance of fertility preservation options, such as oocyte cryopreservation and uterine transplantation, is a burgeoning area of research and clinical practice, highlighting the need for fertility-related discussions and innovations.<sup>3,4</sup>

### 5. Psychosocial and Quality of Life Considerations:

The psychosocial impact of RKH is a dimension of paramount relevance, as individuals grapple with issues related to body image, identity, and sexuality. Therefore, exploring the psychological implications and the development of supportive, patient-centered care strategies are of utmost importance in addressing these non-negligible concerns.<sup>4,5</sup>

### 6. Rare Disease and Advocacy:

Given its rarity, RKH falls under the category of rare diseases. The relevance here extends to the broader context of rare disease advocacy, with RKH serving as an exemplar of the unique challenges faced by individuals living with such conditions, including difficulties in diagnosis, limited

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therapeutic options, and the need for increased awareness and support.<sup>4,5</sup>

In summary, the relevance of Rokitansky-Küster-Hauser Syndrome is multifaceted and far-reaching. It encompasses clinical, diagnostic, genetic, reproductive, psychosocial, and advocacy dimensions. Understanding and addressing these facets is not only pertinent for healthcare professionals but also for the affected individuals and their families, reinforcing the imperative of comprehensive research, multidisciplinary care, and ongoing advocacy to improve the lives of those living with RKH.<sup>4,5</sup>

### CLINICAL MANIFESTATIONS

Rokitansky-Küster-Hauser Syndrome (RKH), also referred to as Müllerian agenesis, is a complex and enigmatic congenital disorder characterized by a distinctive array of clinical manifestations that span across multiple organ systems. Understanding the intricate clinical landscape of RKH is essential for healthcare professionals, researchers, and affected individuals alike. This article provides an exhaustive exploration of the clinical manifestations associated with RKH, encompassing gynecological, urological, skeletal, and psychological aspects.<sup>5,6</sup>

#### Gynecological Manifestations:

**Primary Amenorrhea:** One of the hallmark clinical features of RKH is primary amenorrhea, the absence of menstruation, which typically becomes evident at the expected time of menarche. This absence stems from the underdevelopment or complete absence of the uterus and the upper two-thirds of the vagina.<sup>5,6</sup>

**Normal Secondary Sexual Characteristics:** Despite the absence of a functional uterus, individuals with RKH usually exhibit normal secondary sexual characteristics, including the development of breasts and the presence of pubic and axillary hair.

#### Urological Manifestations:<sup>5,6</sup>

**Renal Anomalies:** A substantial percentage of individuals with RKH may present with urological abnormalities, such as renal agenesis, horseshoe kidneys, and urinary tract malformations. These renal anomalies can necessitate close monitoring and, in some cases, surgical interventions.<sup>5,6</sup>

#### Skeletal Manifestations:

**Skeletal Abnormalities:** RKH is occasionally associated with skeletal anomalies, particularly involving the spine. These may include scoliosis, vertebral anomalies, and other musculoskeletal irregularities. Appropriate evaluation and management of these skeletal manifestations are essential to ensure optimal quality of life.<sup>5,6</sup>

#### Psychological and Psychosocial Manifestations:

**Psychological Impact:** Living with RKH can pose significant psychological challenges, including body image concerns, feelings of difference or isolation, and distress related to the absence of reproductive organs. Psychosocial support and

counseling are vital to address these emotional and mental health aspects.

**Sexuality and Relationships:** The absence of a functional vagina can affect sexual function and may pose challenges in intimate relationships. Addressing these issues and exploring potential therapeutic options are crucial for improving the overall quality of life for individuals with RKH.<sup>5,6</sup>

#### Fertility and Reproductive Considerations:

**Infertility:** RKH typically results in infertility due to the absence of a uterus, which precludes natural conception. Assisted reproductive technologies, such as in vitro fertilization (IVF) with gestational surrogacy, represent potential avenues for individuals with RKH who desire biological children.<sup>5,6</sup>

#### Associated Conditions:

**Endocrine Abnormalities:** Some individuals with RKH may exhibit endocrine abnormalities, such as polycystic ovary syndrome (PCOS) or congenital adrenal hyperplasia, which require appropriate management.<sup>5,6</sup>

**Cardiac Abnormalities:** Rarely, cardiac anomalies may be associated with RKH, emphasizing the need for thorough medical evaluations.

In conclusion, the clinical manifestations of Rokitansky-Küster-Hauser Syndrome are complex and multifaceted, involving gynecological, urological, skeletal, psychological, and reproductive dimensions. Recognizing and addressing these manifestations is essential for healthcare professionals to provide comprehensive care and support to individuals living with RKH.<sup>7,8</sup>

Moreover, fostering awareness and understanding of the syndrome among affected individuals and their families is crucial for enhancing their overall well-being and quality of life.<sup>7,8</sup>

### DIAGNOSIS

The diagnosis of Rokitansky-Küster-Hauser Syndrome (RKH), a rare and complex congenital disorder characterized by the underdevelopment or complete absence of the müllerian duct structures in phenotypic females, presents a multifaceted challenge that requires a thorough and interdisciplinary approach. Accurate and timely diagnosis is pivotal in order to address the physical, psychological, and reproductive implications associated with RKH. This comprehensive article delves into the intricacies of diagnosing RKH, encompassing clinical evaluation, imaging techniques, and genetic assessments.<sup>7,8,9</sup>

### CLINICAL EVALUATION:

The clinical assessment plays a fundamental role in the initial evaluation of individuals suspected of having RKH. A comprehensive medical history, including age of menarche, presence of primary amenorrhea, and family history, is gathered. Gynecological examination reveals the absence or

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underdevelopment of the uterus and upper two-thirds of the vagina, as well as the presence of secondary sexual characteristics. Differentiating between RKH and other causes of primary amenorrhea, such as imperforate hymen or androgen insensitivity syndrome, is a crucial aspect of clinical evaluation.<sup>10,11</sup>

### IMAGING TECHNIQUES:

**Pelvic Ultrasound:** Transabdominal and transvaginal pelvic ultrasound is often employed to visualize the pelvic anatomy. In RKH, these ultrasounds reveal the absence or hypoplasia of the uterus and the upper vaginal segment. The presence of normal-sized ovaries is a significant finding in RKH.<sup>10,11</sup>

**Magnetic Resonance Imaging (MRI):** Pelvic MRI is increasingly utilized for its superior soft tissue resolution, which aids in confirming the absence or underdevelopment of the müllerian structures and the presence of other pelvic anomalies. Additionally, MRI can provide valuable information for preoperative planning, especially in cases where surgical intervention is being considered.<sup>10,11</sup>

### Genetic Assessment:

Emerging evidence supports the relevance of genetic evaluation in diagnosing RKH and uncovering potential underlying genetic factors. While a genetic basis for RKH remains incompletely understood, genetic testing, such as whole exome sequencing or targeted gene analysis, may reveal mutations or variants in candidate genes like WNT4 or HOXA, which are associated with müllerian duct development. Genetic counseling is essential in such cases, not only for diagnostic purposes but also for guiding family planning decisions.<sup>10,11</sup>

### Psychosocial Evaluation:

Psychological and psychosocial assessments are a crucial aspect of the diagnostic process, as individuals diagnosed with RKH may require support and counseling to address the emotional and mental health aspects of living with the condition. These evaluations help individuals cope with body image concerns, sexuality, and the unique challenges associated with RKH.<sup>10,11</sup>

### Differential Diagnosis:

It is imperative for clinicians to differentiate RKH from other causes of primary amenorrhea and müllerian anomalies, such as androgen insensitivity syndrome, congenital adrenal hyperplasia, and imperforate hymen. A precise diagnosis is essential to ensure appropriate management and support tailored to the specific condition.<sup>11</sup>

In conclusion, the diagnosis of Rokitansky-Küster-Hauser Syndrome is a multifaceted process that encompasses clinical evaluation, advanced imaging techniques, genetic assessment, and psychosocial considerations. Achieving an accurate diagnosis is not only pivotal for understanding the individual's medical condition but also for providing comprehensive care and support to address the multifaceted

physical, psychological, and reproductive implications associated with RKH.<sup>12</sup>

### TREATMENT STRATEGIES

Rokitansky-Küster-Hauser Syndrome (RKH), a rare congenital disorder characterized by the absence or underdevelopment of müllerian duct structures in phenotypic females, necessitates a multi-pronged, multidisciplinary approach to address its complex physical, psychological, and reproductive ramifications. The management of RKH is contingent upon individual patient preferences, age, and the specific clinical presentation. This comprehensive article delves into the intricate landscape of treatment options and strategies available to individuals living with RKH, encompassing gynecological, surgical, psychological, and reproductive dimensions.<sup>12</sup>

#### Gynecological Management:

**Vaginal Dilators:** For individuals with RKH who desire sexual intercourse or the ability to undergo vaginal examinations, a gradual approach to vaginal dilation is often recommended. Vaginal dilators can facilitate the expansion of the vaginal canal over time, improving both sexual function and quality of life.<sup>12</sup>

**Vaginal Construct:** In cases where dilators do not provide the desired outcome or are not well-tolerated, the creation of a neovagina via surgical techniques, such as the McIndoe or Vecchiotti methods, can be considered. These procedures involve the surgical formation of a functional vaginal canal.<sup>12</sup>

#### Surgical Interventions:

**Neovaginal Construction:** In instances where the absence of a vaginal canal poses significant physical or psychological distress, surgical neovaginal construction represents a viable option. These surgical techniques aim to create a functional vaginal canal using various approaches, including laparoscopic, abdominal, or perineal methods.<sup>12</sup>

**Uterine Transplantation:** For individuals with RKH who aspire to experience pregnancy and childbirth, uterine transplantation has emerged as a groundbreaking and experimental intervention. This complex procedure involves the transplantation of a donor uterus to enable gestation.<sup>12</sup>

#### Psychological Support:

**Psychological Counseling:** The psychological aspect of living with RKH is paramount, as individuals may grapple with body image concerns, issues related to sexuality, and the emotional implications of primary amenorrhea and infertility. Psychological counseling, support groups, and therapy can help individuals navigate these emotional challenges.<sup>12</sup>

#### Reproductive Considerations:

**Assisted Reproductive Technologies (ART):** In cases where pregnancy is desired, despite the absence of a uterus, gestational surrogacy represents a viable option. ART techniques, such as in vitro fertilization (IVF), allow for the

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use of the individual's eggs, fertilization with a partner's or donor sperm, and the transfer of the resulting embryos to a gestational carrier.<sup>12</sup>

**Fertility Preservation:** Prior to undergoing any interventions that may impact ovarian function, it is imperative to consider fertility preservation methods, such as oocyte cryopreservation, to preserve the potential for biological motherhood.<sup>12</sup>

### Long-Term Follow-Up:

Regular long-term follow-up is essential to monitor the physical and psychological well-being of individuals with RKH. It allows for the identification and management of potential complications, such as urinary tract abnormalities, and ensures the ongoing appropriateness of treatment strategies in light of evolving personal circumstances and preferences.<sup>13</sup>

### Patient-Centered Care:

The management of RKH should always be tailored to the individual's unique needs and desires. Patient-centered care, including shared decision-making with healthcare providers, is pivotal in the selection of appropriate treatment strategies and the ongoing support of individuals living with RKH.<sup>13</sup>

In conclusion, the treatment landscape for Rokitansky-Küster-Hauser Syndrome is multifaceted and highly individualized, encompassing gynecological, surgical, psychological, and reproductive dimensions. Achieving optimal patient outcomes necessitates a comprehensive, multidisciplinary approach, prioritizing the physical and psychological well-being of individuals affected by RKH. Moreover, ongoing research and innovation continue to expand the horizons of treatment options, offering hope and improved quality of life for those living with this complex congenital condition.<sup>13</sup>

## CONCLUSIONS

The journey through the multifaceted landscape of Rokitansky-Küster-Hauser Syndrome (RKH) has unveiled a spectrum of complexities, from the clinical manifestations to the diagnostic intricacies, and from the diverse treatment modalities to the profound psychosocial dimensions. Through this comprehensive exploration, several key takeaways emerge, shedding light on the path forward for understanding and managing this rare and enigmatic condition.

1. **Multidisciplinary Collaboration is Paramount:** RKH underscores the compelling need for a multidisciplinary approach to healthcare. The diagnosis and management of RKH require the coordinated efforts of gynecologists, urologists, geneticists, radiologists, and psychologists, among others. The synergy among these specialties is essential to providing comprehensive care, addressing the physical and emotional aspects, and ensuring the well-being of affected individuals.

2. **A Spectrum of Treatment Options:** The treatment options for RKH are as diverse as the clinical presentations themselves. From vaginal dilators to surgical neovaginal construction, and from fertility preservation to gestational surrogacy, the array of choices empowers individuals with RKH to make decisions that align with their personal preferences and aspirations. The individual's voice, desires, and values must be central in the decision-making process.

3. **Fertility Preservation and Reproductive Freedom:** Advances in assisted reproductive technologies and uterine transplantation have opened new avenues for individuals with RKH who aspire to become parents. Fertility preservation options, such as oocyte cryopreservation, provide the opportunity for biological motherhood, while the potential for uterine transplantation offers hope for gestation. These groundbreaking developments signify a profound shift in the landscape of reproductive possibilities for those affected by RKH.

4. **The Importance of Psychological Support:** The emotional aspects of living with RKH cannot be understated. The absence of a functional uterus, primary amenorrhea, and fertility challenges can elicit a range of psychological responses. Psychological support, including counseling and therapy, plays an instrumental role in addressing body image concerns, issues related to sexuality, and overall psychosocial well-being.

5. **Rare Disease Advocacy:** RKH's status as a rare condition underscores the broader context of rare disease advocacy. The challenges in diagnosis, limited therapeutic options, and the need for increased awareness underscore the importance of advocacy efforts, research, and support for individuals living with RKH and other rare diseases.

6. **Ongoing Research and Innovation:** The future of RKH management holds promise, with ongoing research endeavors seeking to unravel the genetic underpinnings and further refine diagnostic and treatment strategies. The landscape is dynamic, and the relentless pursuit of knowledge and innovation continues to expand the horizons of care for individuals with RKH.

In the grand tapestry of medicine, Rokitansky-Küster-Hauser Syndrome represents a unique and intricate thread. Its rarity, complexity, and multidimensional impact necessitate not only the compassionate care of healthcare providers but also the resilience and determination of individuals and their families. As we move forward, it is imperative to foster greater awareness, understanding, and support for those living with RKH, and to continue the quest for improved diagnostic, therapeutic, and psychological interventions.

RKH is not merely a medical condition; it is a testament to the resilience of the human spirit and the capacity of science and medicine to adapt and evolve. With the spotlight shining brighter on RKH, we navigate the path forward with compassion, innovation, and unwavering dedication to

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improving the lives of those affected by this rare and enigmatic syndrome.

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