

## Cutis Verticis Gyrata and Quality of Life: Clinical Report of 13 Cases

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### ABSTRACT

**Background:** The quality of life of patients with Cutis Verticis Gyrata (CVG) has not been previously investigated in the literature.

**Objective:** Description of features and evaluation of quality of life of CVG patients.

**Materials and methods:** We employed a mixed methods study. We retrospectively collected data from the clinical database from 2012 to 2020. We selected 17 clinically diagnosed CVG patients and prospectively evaluated the quality of life of these CVG patients.

**Results:** Out of 13 patients, 11 patients were in between the 14-47 age group. The majority of them were male (77%), Caucasian (54%), had no significant family history (92.3%), and had the age of onset below < 30 years (69.2%). The majority of the patients had a secondary form of CVG (54%) followed by a primary essential form of CVG (46.1%). The CVG had a small effect on the majority of patient's quality of life (77%), and a majority of patients were not taking any treatment (69.2%)

**Conclusion:** In our study, the CVG was shown to have a small effect on the patient's quality of life.

**Keywords:** Acromegaly, Cutis verticis gyrata, DLQI, Quality of life, Skin diseases.

### INTRODUCTION

Cutis Verticis Gyrata (CVG) is a rare benign cutaneous disorder characterized by the scalp's convoluted folds and deep furrows that mimic cerebral sulci and gyri<sup>(1)</sup>. The primary essential CVG is not linked to any other diseases, whereas the primary non-essential form is linked to mental deficiency, neurological diseases, and ophthalmological disorders<sup>(2)</sup>. Secondary CVG can be caused by many factors, including inflammatory dermatoses, systemic diseases, infiltrates, and cutaneous neoplasms<sup>(3)</sup>.

Clinical, pathological, and radiological features of cutis verticis gyrata patients were reported in many studies<sup>(2-6)</sup>. As far as we know, patients' quality of life with CVG has not been previously investigated in the literature.

Dermatology Life Quality Index (DLQI) scale helps measure how much the skin disease has affected patients' lives suffering from dermatological disorders<sup>(7)</sup>.

So, we aimed to evaluate the patient's quality of life of different forms of cutis verticis gyrata using the Dermatology Life Quality Index (DLQI) scale.

### PATIENTS AND METHODS

A mixed-methods study was carried out using a clinical database in Sabouraud Center – Saint Louis University Hospital, Paris. We found 17 clinically diagnosed CVG cases from 120000 patients in the database as it is a rare disease. Out of 17 patients, we were unable to reach four patients.

So, we presented 13 cases of CVG. The following information was collected from the patient's case sheet: age, gender, ethnicity, family history, age of

onset, type of CVG, risk factors, complications, presence of depression or anxiety, treatment, and disease course. The information on Dermatology Life Quality Index was collected from the patient.

### Ethical consideration

An approval of the study was obtained from Sabouraud Center – Saint Louis University Hospital, (Paris, France) Academic and Ethical committee. The aim of the study was clearly explained to the 13 patients, and written informed consent was collected from them. This work has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for studies involving humans.

### Statistical analysis

The collected data were coded, processed and analyzed using the SPSS (Statistical Package for the Social Sciences) version 22 for Windows® (IBM SPSS Inc, Chicago, IL, USA).

### RESULTS

As shown in table (1), 11 patients were in between the 14-47 age group. The majority of them were male (77%), Caucasian (54%), had no significant family history (92.3%), and had the age of onset below < 30 years (69.2%). The majority of the patients had a secondary form of CVG (54%) followed by a primary essential form of CVG (46.1%), had no complications (69.2%), and no depression or anxiety (69.2%).



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**Table (1):** Sociodemographic and clinical characteristics of the patients (n=13).

Patient	Gender	Ethnicity	Age (years)	Age of Onset (years)	Category	Dermatological Complications	Depression or Anxiety	Secondary to
1	M	Middle Eastern	16	10	Secondary	No	No	Neurofibroma
2	M	African	19	18	Primary Essential	No	No	N/A
3	M	Caucasian	34	24	Secondary	No	Yes	Dissecting Cellulitis
4	F	Middle Eastern	22	19	Primary Essential	Folliculitis	Yes	N/A
5	F	Caucasian	67	Since birth	Secondary	No	Yes	Cerebriform Intradermal Nevus
6	M	Caucasian	25	23	Primary Essential	Inflammation and Folliculitis	No	N/A
7	M	African	38	33	Secondary	No	No	Lipoedema
8	M	African	31	22	Primary Essential	No	No	N/A
9	M†	Caucasian	29	17	Secondary	No	No	Folliculitis Fibrosis
10	M	Caucasian	42	25	Primary Essential	Bleeding and Irritation	No	N/A
11	F	Caucasian	14	12	Primary Essential	No	No	N/A
12	M	Caucasian	44	37	Secondary	No	Yes	Chronic Folliculitis
13	M	Middle Eastern	51	40	Secondary	Inflammation	No	Acromegaly

†M= ONLY patient with significant family history

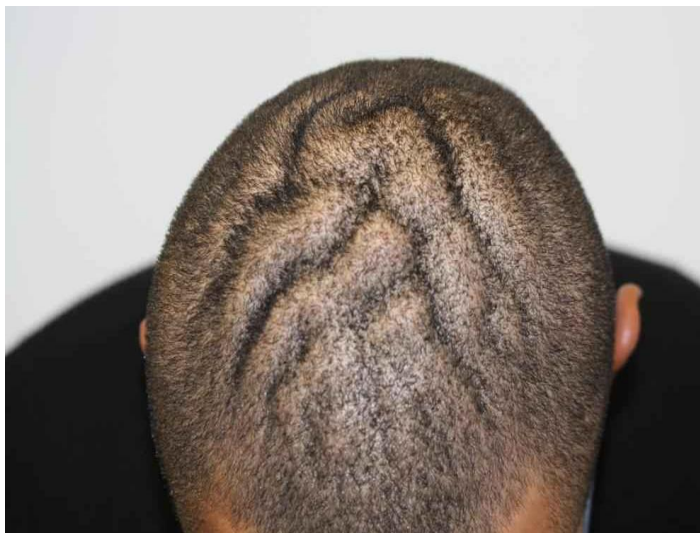
The majority of the patients were having a stable course of disease (69.2%). The CVG had a small effect on the majority of patient's quality of life (77%), and a majority of patients are not taking any treatment (69.2%) (Table 2).

**Table (2):** Quality of life of the patients (n=13).

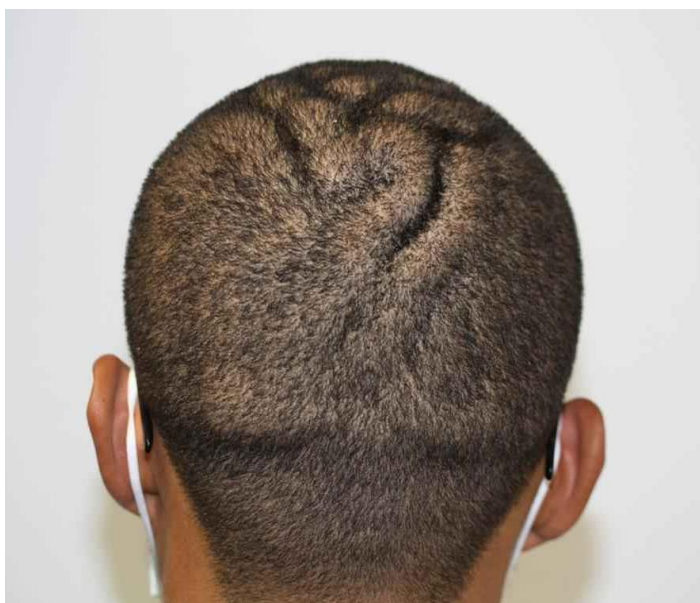
Patient	Gender	Disease Course	DLQI Score	Effect on QoL	Treatment
1	M	Small Improvement	4	Small Effect	Not taking any treatment
2	M	Stable	4	Small Effect	Not taking any treatment
3	M	Small Improvement	6	Moderate Effect	Not taking any treatment
4	F	Stable	6	Moderate Effect	Doxycycline. Antiseptic foaming solution, Antifungal shampoo
5	F	Good Improvement	6	Moderate Effect	2 surgeries
6	M	Stable	4	Small Effect	Doxycycline; Antifungal shampoo
7	M	Stable	4	Small Effect	Not taking any treatment
8	M	Stable	4	Small Effect	Not taking any treatment
9	M	Stable	6	Moderate Effect	Not taking any treatment
10	M	Stable	4	Small Effect	Not taking any treatment
11	F	Stable	4	Small Effect	Not taking any treatment
12	M	Small Improvement	4	Small Effect	Not taking any treatment
13	M	Stable	4	Small Effect	Antifungal shampoo

DLQI: Dermatology Life Quality Index, QoL: Quality of life

Typical features of cutaneous scalp hypertrophy due to CVG are shown in figures 1 and 2.



**Fig. (1):** Scalp hypertrophy in parietal region of Cutis Verticis Gyrate (CVG) patient.



**Fig. (2):** Cutaneous hypertrophy with cerebriform aspect of scalp. Anteroposterior of disposition in parietal region

## DISCUSSION

Men are more affected in our study. Cutis verticis gyrata<sup>(1,8)</sup>, especially primary essential form occurs predominantly in men<sup>(9)</sup>. Out of 13 patients, only one patient had a significant family history in our study. There are only two familial occurrences of CVG in literature<sup>(10)</sup>.

Majority of the studies reported insignificant family history of CVG<sup>(2, 9, 11)</sup>. We observed that the majority of patient's age of onset of CVG was below 30 years. The majority of primary cases appeared after puberty and, in 90% of instances, before 30<sup>(12)</sup>. We observed a congenital CVG secondary to cerebriform intradermal nevus<sup>(13)</sup>.

Secondary form of CVG is high in our study. To the best of our knowledge, this is the first study to report CVG secondary to neurofibroma, folliculitis

fibrosis, dissecting cellulitis, and lipoedema. Previous studies reported CVG secondary to acromegaly<sup>(14, 15)</sup>, cerebriform intradermal nevus<sup>(2, 3)</sup>. We were unable to find the primary non-essential form of CVG, which is rare and exceptional.

Of 13 patients, CVG had a small effect on the quality of life of most patients (n=9) in our study. The disease course of improvement also differed among the subjects having a small effect and moderate effect on the quality of life. The quality of life differed among patients taking treatment vs not taking treatment. These differences were may be due to differences in genetic make-up or the severity of the disease.

The quality of care for CVG treatment involves surgical intervention for cosmetic purposes, and in the case of secondary CVG, treatment is directed toward resolving the underlying systemic disease<sup>(16)</sup>. Surgical treatment may enhance the clinical characteristic, but the aesthetic element may impair patients' quality of life in patients with cutis verticis gyrata<sup>(9)</sup>.

In one study facial cutis verticis gyrata distorted the quality of life of four patients<sup>(17)</sup>. However, after direct excision of large and thickened gyri form folds, all patients reported a significant and lasting improvement in their quality of life in self-esteem, and social life<sup>(17)</sup>. The treatment choices for CVG are limited and frequently require invasive procedures. Hyaluronidase injections offer an intriguing alternative that can significantly enhance a patient's quality of life. Six injections of 150 units of hyaluronidase every six weeks may show promising improvement in the condition and thus the quality of life<sup>(16)</sup>.

The influence of underlying systemic diseases must be considered on the patient's quality of life. For example, the majority of individuals with acromegaly experience depression, sleep difficulties, impaired body image, diabetes mellitus, and arterial hypertension, all of which contribute to poor quality of life and increased morbidity and mortality as a result of prolonged exposure to elevated growth hormone secretion<sup>(18)</sup>.

Chronic nature and low mortality may underestimate the importance of skin disease. However, skin diseases cause physical and psychological impairment and affect self-image. Unfavourable self-image leads to anxiety, depression, anger, fear, and social maladjustment<sup>(19)</sup>. So, evaluating the quality-of-life patients with cutis verticis gyrata may help determine and evaluate treatment modalities.

## CONCLUSION

We present 13 patients with different forms of cutis verticis gyrata and their quality of life. We want to highlight the importance of quality-of-life measurements in cutis verticis gyrata patients.

It helps to evaluate the impact of the treatment modalities and helps to choose better treatment in multidisciplinary team when needed.

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