

Craniopharyngioma

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References

The background of the slide is a photograph of a vast, calm ocean under a blue sky with wispy white clouds. The water is a deep blue, and the horizon line is visible in the distance. The overall color palette is dominated by various shades of blue.

Introduction

- Craniopharyngiomas
 - Benign, epithelial neoplasm
 - Sellar and parasellar
 - Arise from neoplastic transformation of ectoderm derived epithelial remnants of Rathke's pouch and the craniopharyngeal duct

Passion of the Craniopharyngioma

- Cushing commented that craniopharyngiomas were the “most forbidding of the intracranial tumors”
- Rutka commented that “there is perhaps no other brain tumor that evokes more passion, emotion, and controversy as craniopharyngioma”

Passion of the Craniopharyngioma

- Therapeutic goals are the cure of disease with functional preservation and restoration.
- This is often a widely debated topic with controversy including extent of surgical resection, surgical approach, and the use of adjuvant therapy

History

- In 1838, Rathke described the contribution of the ectodermal stomadeal diverticulum in the development of the anterior hypophysis
- In 1857, Zenker identified masses of cells resembling squamous epithelium along the pars distalis and pars tuberalis of the pituitary
- In 1904, Erdheim ascribed the origins of this tumor to vestiges of an incompletely involuted hypophyseal pharyngeal duct

History

- Halstead reported in 1910 the first attempted resection at Northwestern University
- In 1932, Cushing introduced the term "craniopharyngioma" noting that "this somewhat cumbersome term has been employed for want of something brief to include kleidoscopic tumors, solid and cystic, which take their origin from epithelial rests ascribable to an imperfect closure of the hypophyseal or craniopharyngeal duct"

History

The background of the slide is a blue-tinted photograph of a vast, calm ocean stretching to a clear horizon. The sky is filled with soft, wispy white clouds. The water's surface shows gentle ripples, and a bright reflection of light is visible on the left side of the horizon.

History

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Epidemiology

- Rare tumors
- Overall incidence 0.13 per 100,000
- 2-5% of all primary intracranial neoplasms
- 5-15% of intracranial tumors in children
- About half of all cases are diagnosed in adults
- Approximately 338 cases of craniopharyngiomas are expected to occur annually in the United States, with 96 occurring in children from 0 to 14 years of age

Epidemiology

- Bimodal age distribution with peaks at ages 5-14 and 50-74
- No gender differences noted
- Unclear if there are any genetic susceptibilities. There have been reports of involvement of chromosome 2 and 12.

Epidemiology

QuickTime™ and a
TIFF (Uncompressed) decompressor
are needed to see this picture.

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Patient Presentation: Child

DeVries et al, 2003

- 36 children with craniopharyngioma
- Presenting symptoms include:
 - headache (51.4%)
 - vomiting (31%)
 - visual disturbances (22.9%)
 - polyuria and/or polydipsia (17.1%)
 - delayed puberty (19.4%)
 - short stature (13.8%)
 - precocious puberty (2.7%)

Patient Presentation: Adult

The background of the slide is a deep blue gradient. The upper portion features a lighter blue sky with wispy white clouds. A thin, bright white horizon line separates the sky from the lower portion, which depicts a vast expanse of blue water with fine, rhythmic ripples. The overall effect is a serene, naturalistic scene.

Pathogenesis

- Review of Pituitary Embryogenesis
 - Primitive streak and node appear on the dorsal aspect of the embryonic disc during 3rd week
 - Median neural groove, flanked by neural folds, fuses in midline forming a tubular structure that disengages from the surface ectoderm
 - A series of evaginations of the neural tube develop producing the pineal, optic stalks, cerebral hemispheres, and infundibulum
 - An ectodermal outgrowth develops from the buccopharyngeal membrane forming Rathke's pouch which extends cephalad toward the infundibulum

Pathogenesis

- Anterior wall of Rathke's pouch forms:
 - pars distalis -> bulk of adenohypophysis
 - Pars tuberalis -> glandular portion of infundibular stalk
- Posterior wall of Rathke's pouch forms:
 - Vestigial pars intermedia
- Stomodeum differentiates into tooth primordia and nonkeratinized squamous epithelium of buccal mucosa
- 12th week: development of sphenoid bone, separates sella from oral cavity

Pathogenesis

- Embryology Images

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Pathogenesis

- Review of Pituitary Anatomy
- 2 main parts posterior, or **neurohypophysis** and anterior, or **adenohypophysis** (the pars distalis, pars intermedia, and pars tuberalis).
- Between the two a specialized vascular (portal) system allows communication from the brain to peripheral endocrine organs and other systems.

Pathogenesis

- Anatomy images

Pathogenesis

- Two primary subtypes
 - Adamantinomatous
 - Papillary

Pathogenesis

- Adamantinomatous Subtype
 - More common form
 - Occurs in children
 - Cysts with turbid, yellow, cholesterol rich fluid
 - Solid portions with clusters of organized palisading columnar cells that form whorls and sheets around loose stellate epithelial cells
 - Desquamated cells form large, pale, eosinophilic masses called WET KERATIN
 - Growth into brain can produce a gliotic capsule with Rosenthal fibers

Pathogenesis

- Histology

Pathogenesis

- Papillary Subtype
 - Occur in adults
 - Solid
 - No calcification, no motor oil
 - Papillae of bland, well-differentiated squamous epithelium growing around a fibrovascular core

Pathogenesis

- Papillary Images

Pathogenesis

- Theories of origin

- (1) Embryogenetic Theory: adamantinomatous CP arise from epithelial remnants of the craniopharyngeal duct or Rathke's pouch derived from stomodeum destined to form tooth primordia
- (2) Metaplastic Theory: papillary tumors arise from metaplasia of squamous epithelial rests that are remnants of that portion of the stomodeum that contributed to the development of the buccal mucosa

Pathogenesis

- FYI: Rathke's Cleft Cyst
 - Non-neoplastic
 - Located between pars distalis and pars intermedia
 - Lined by ciliated cylindrical epithelium
 - Lack solid component

Growth Pattern

- Although histologically benign, craniopharyngiomas can act malignant due to tendency to recur, growth pattern, and proximity to important structures

Growth Patterns

- Hoffman et al, suggest 4 categories of CP growth:
 - (1) Sellar craniopharyngioma
 - (2) Prechiasmatic craniopharyngioma
 - (3) Retrochiasmatic craniopharyngioma
 - (4) Giant craniopharyngioma

Growth Pattern

Wang et al, CNS 2005

- Classification of CP by location of origin
 - (1) Subdiaphragmatic with competent diaphragma sella
 - (2) Subdiaphragmatic with incompetent diaphragma sella
 - (3) Supradiaphragmatic

Growth Pattern

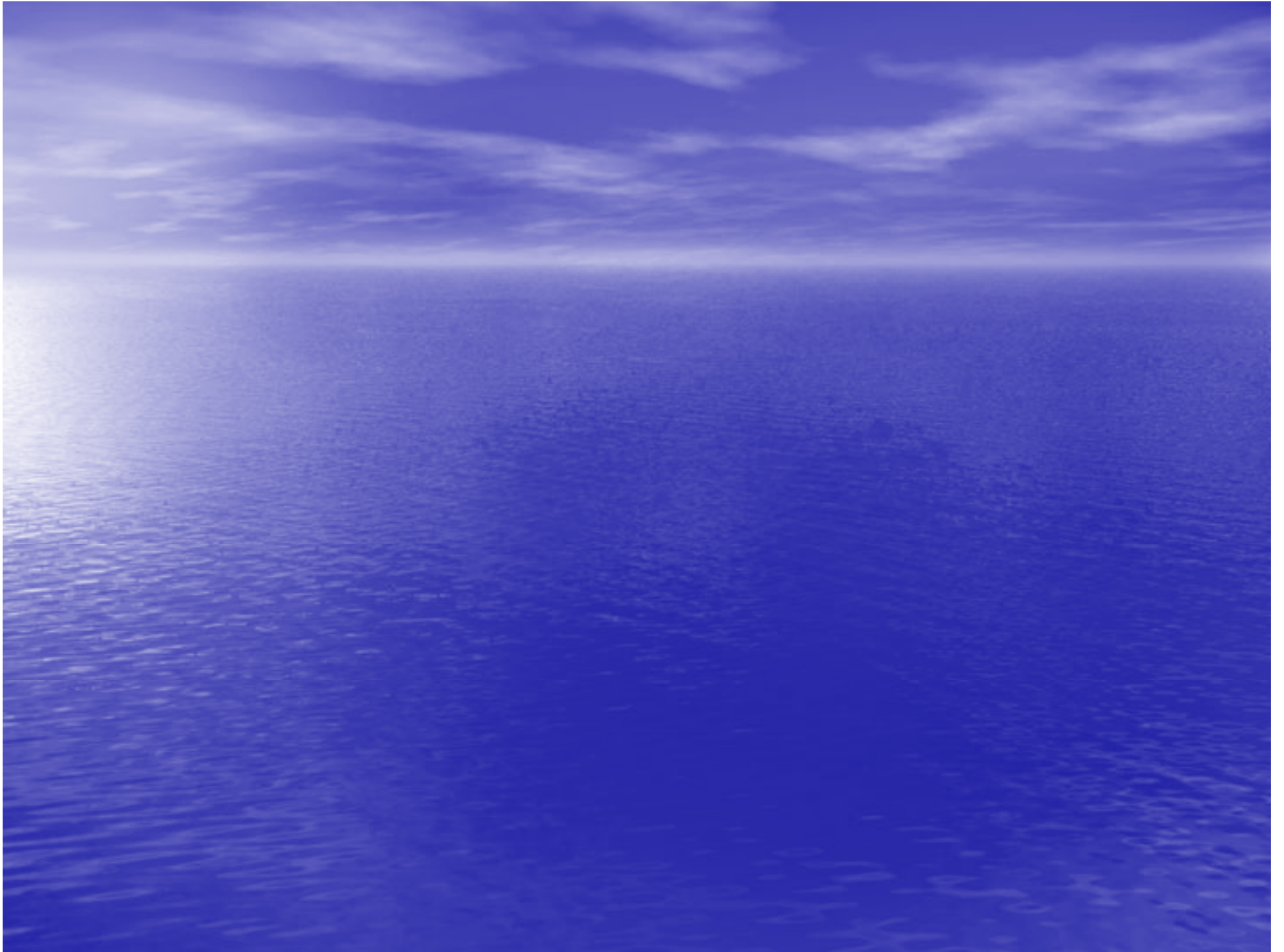
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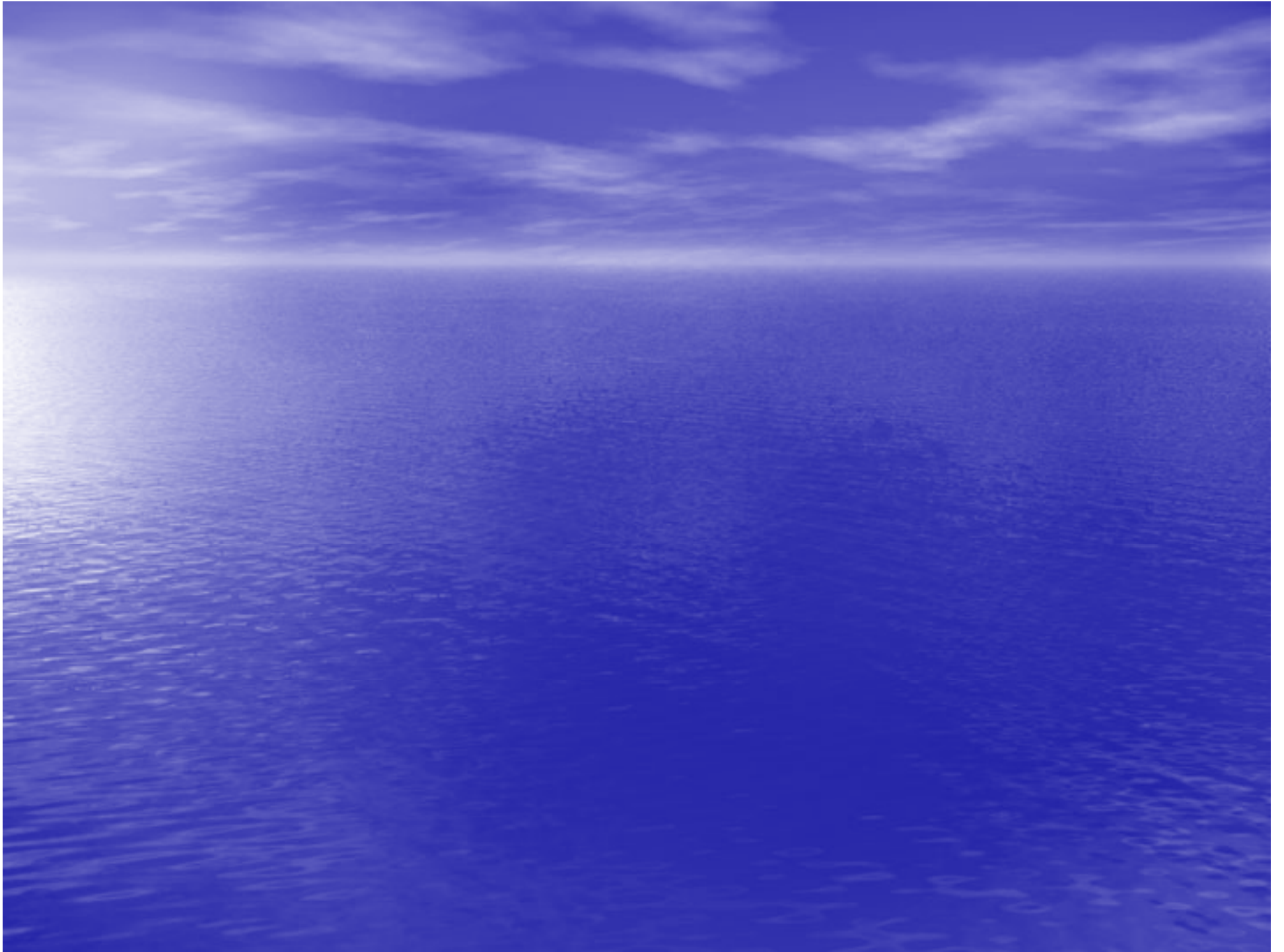
Growth Pattern

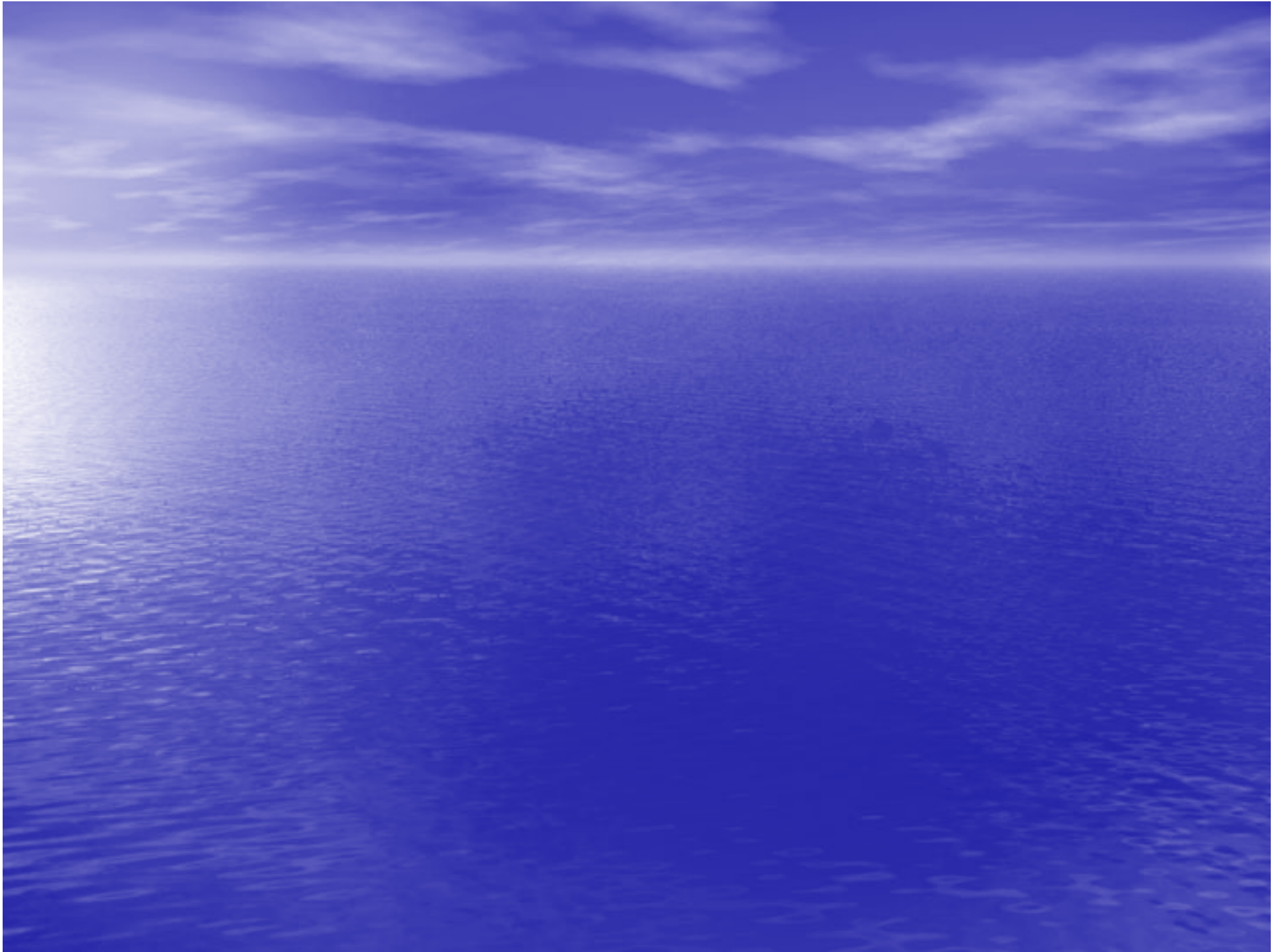
The image features a full-page background of a blue-tinted photograph. The top half shows a sky with wispy, light-colored clouds. The bottom half shows a vast, calm ocean with subtle ripples on its surface. The overall color palette is a range of blues, from light sky blues to deep, dark blues in the water.

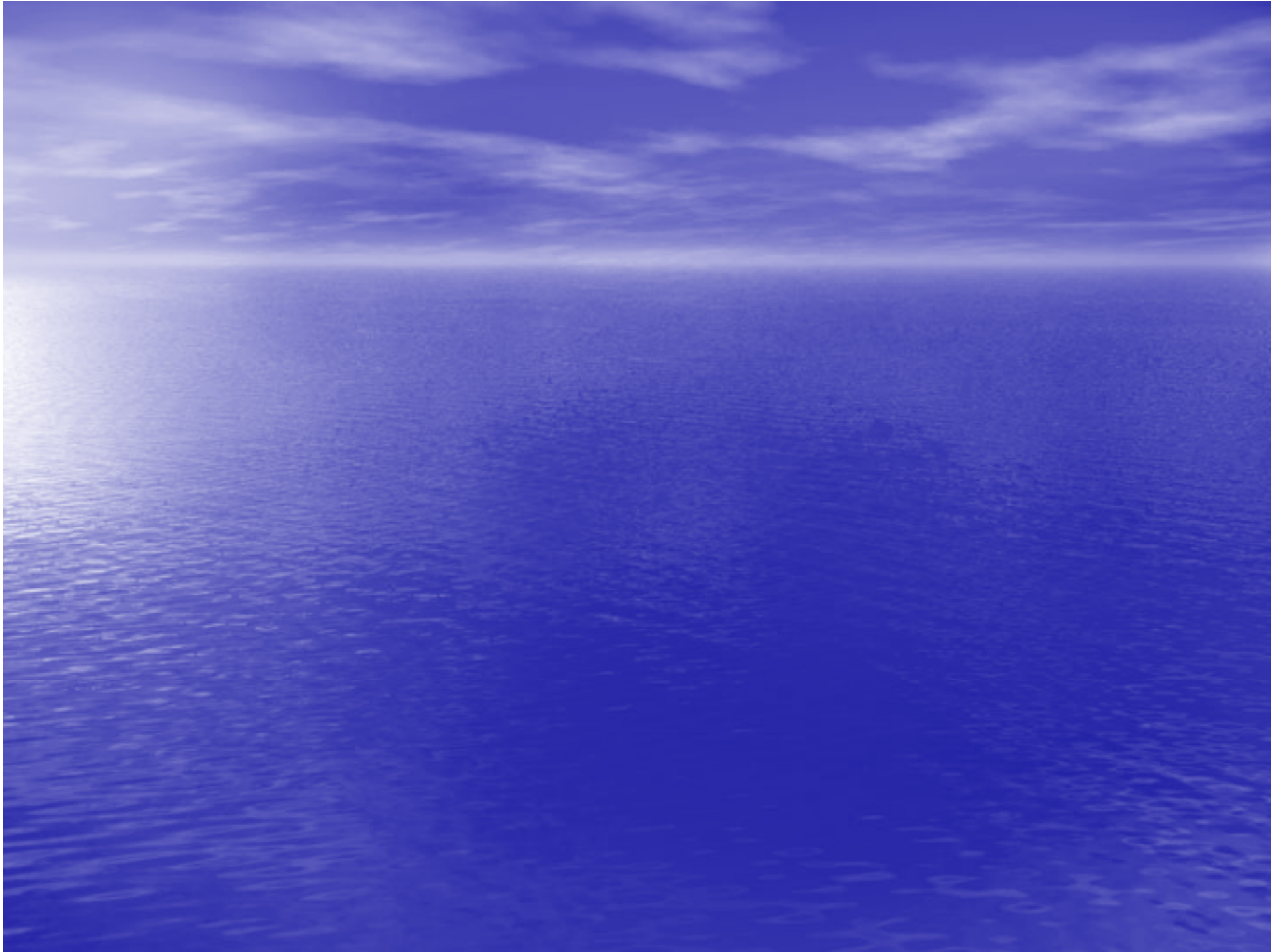
Imaging

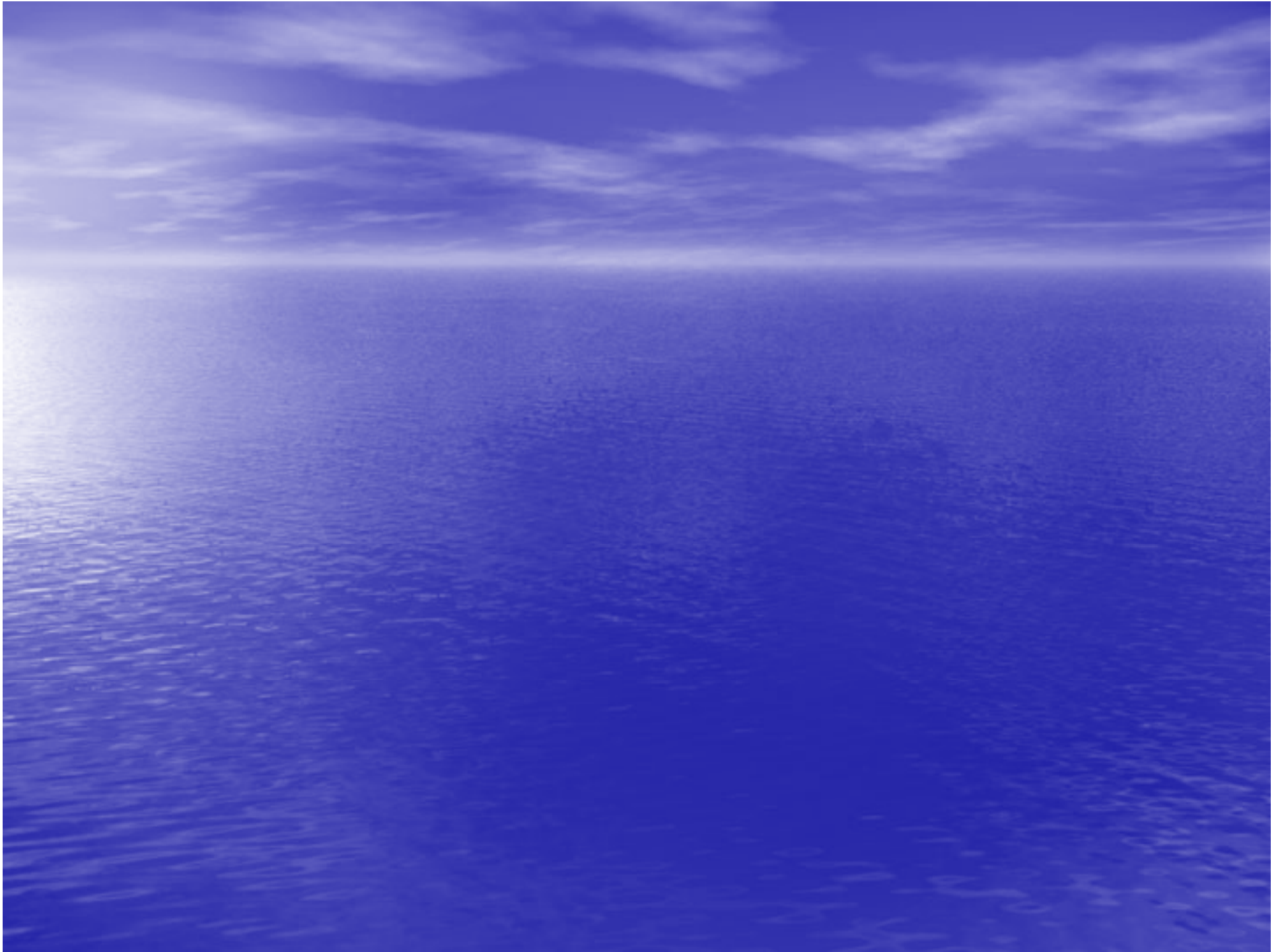
A blue-tinted photograph of a vast ocean under a cloudy sky. The word "Imaging" is written in white text in the upper left corner. The image shows a wide expanse of water meeting a horizon line, with a sky filled with soft, white clouds. The overall color palette is dominated by various shades of blue, from deep navy to light sky blue.

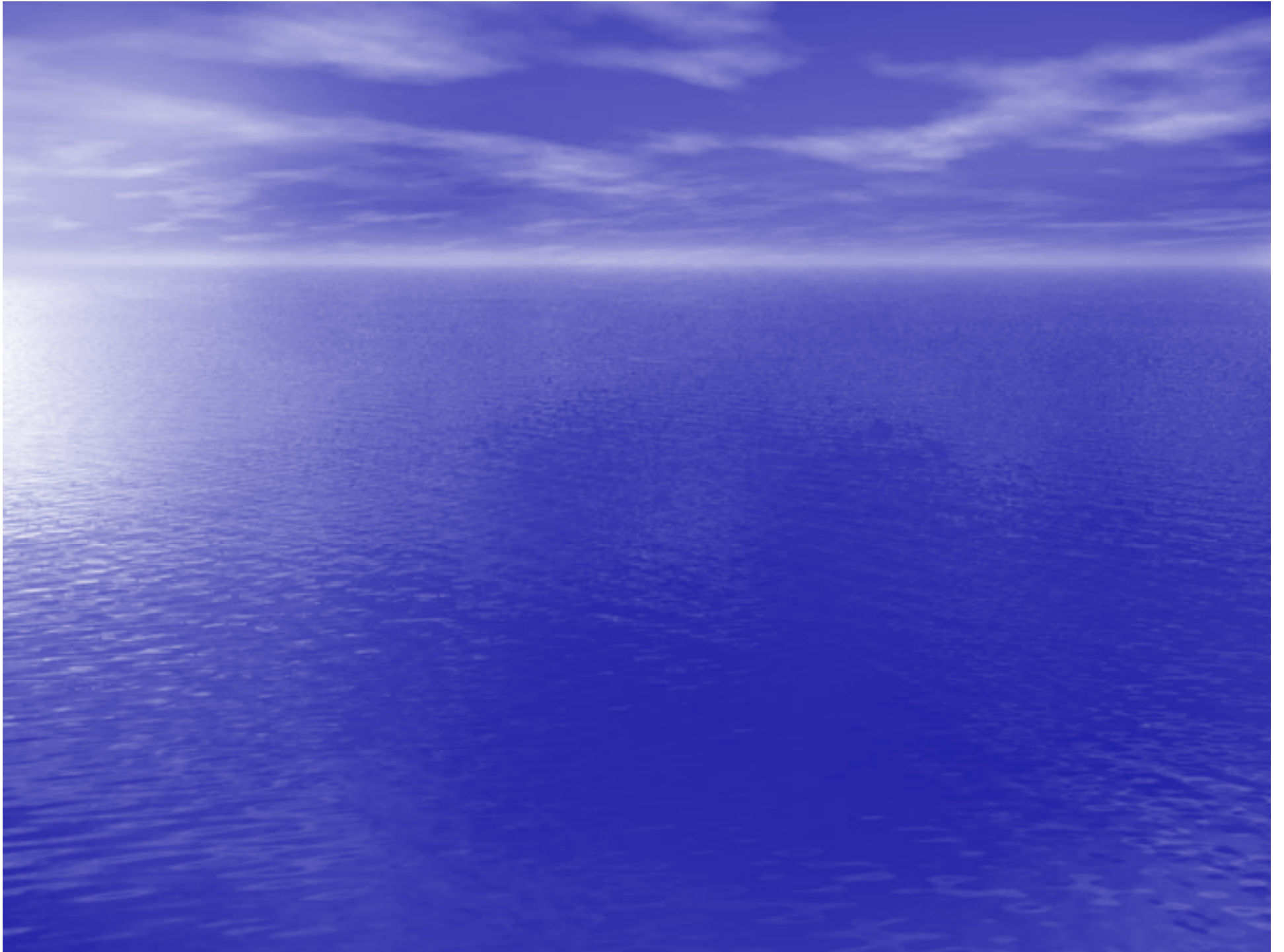






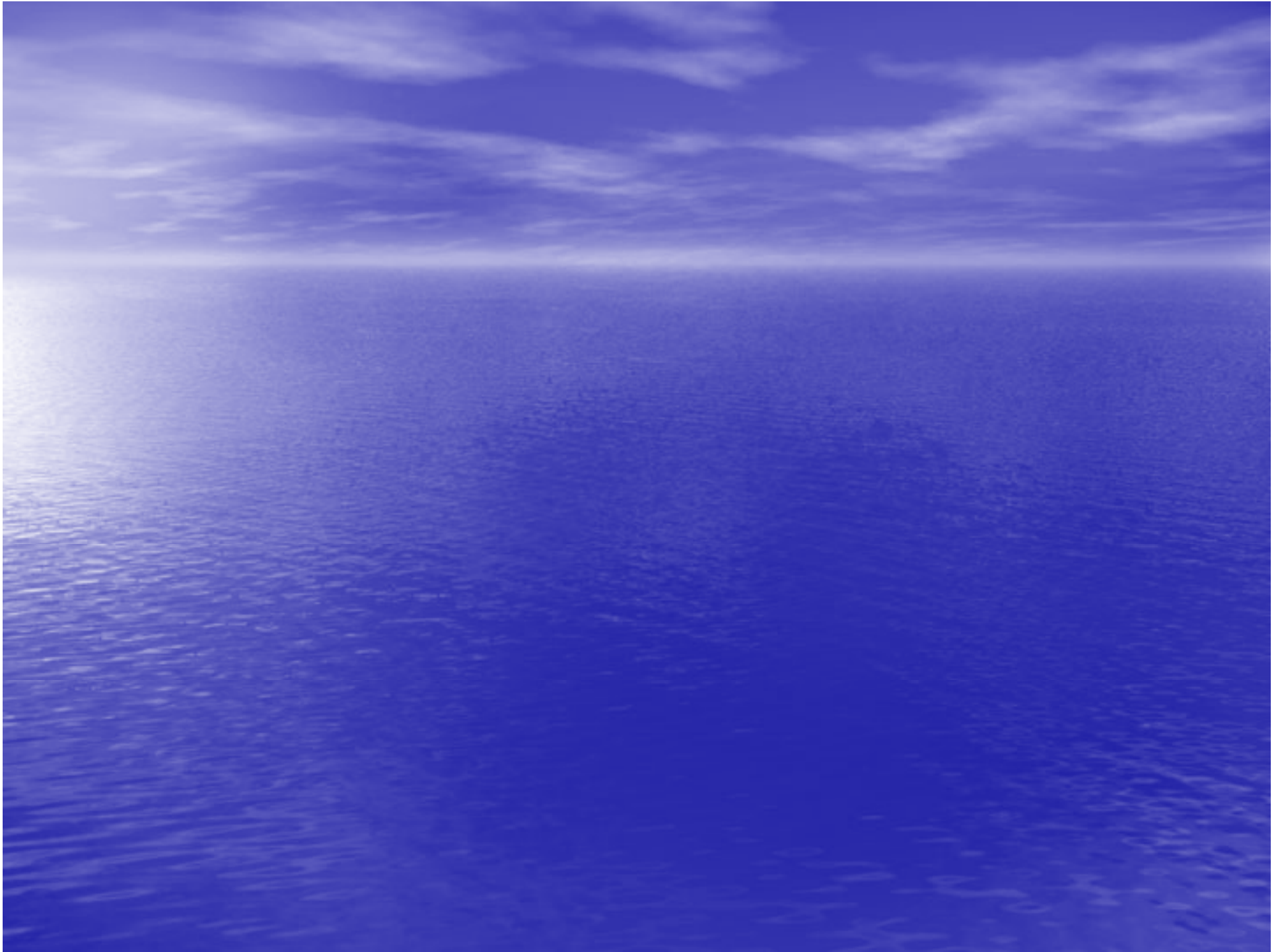


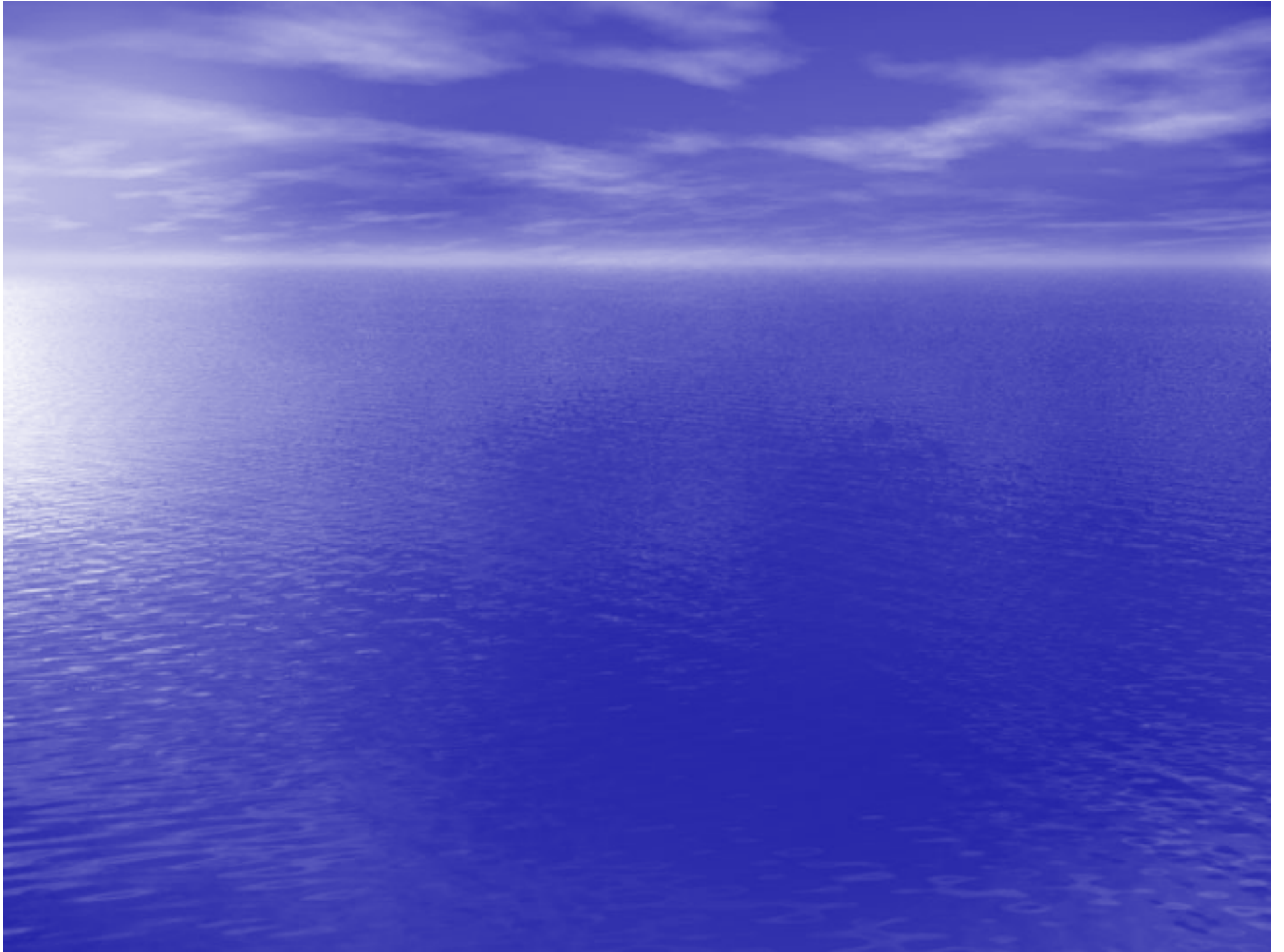


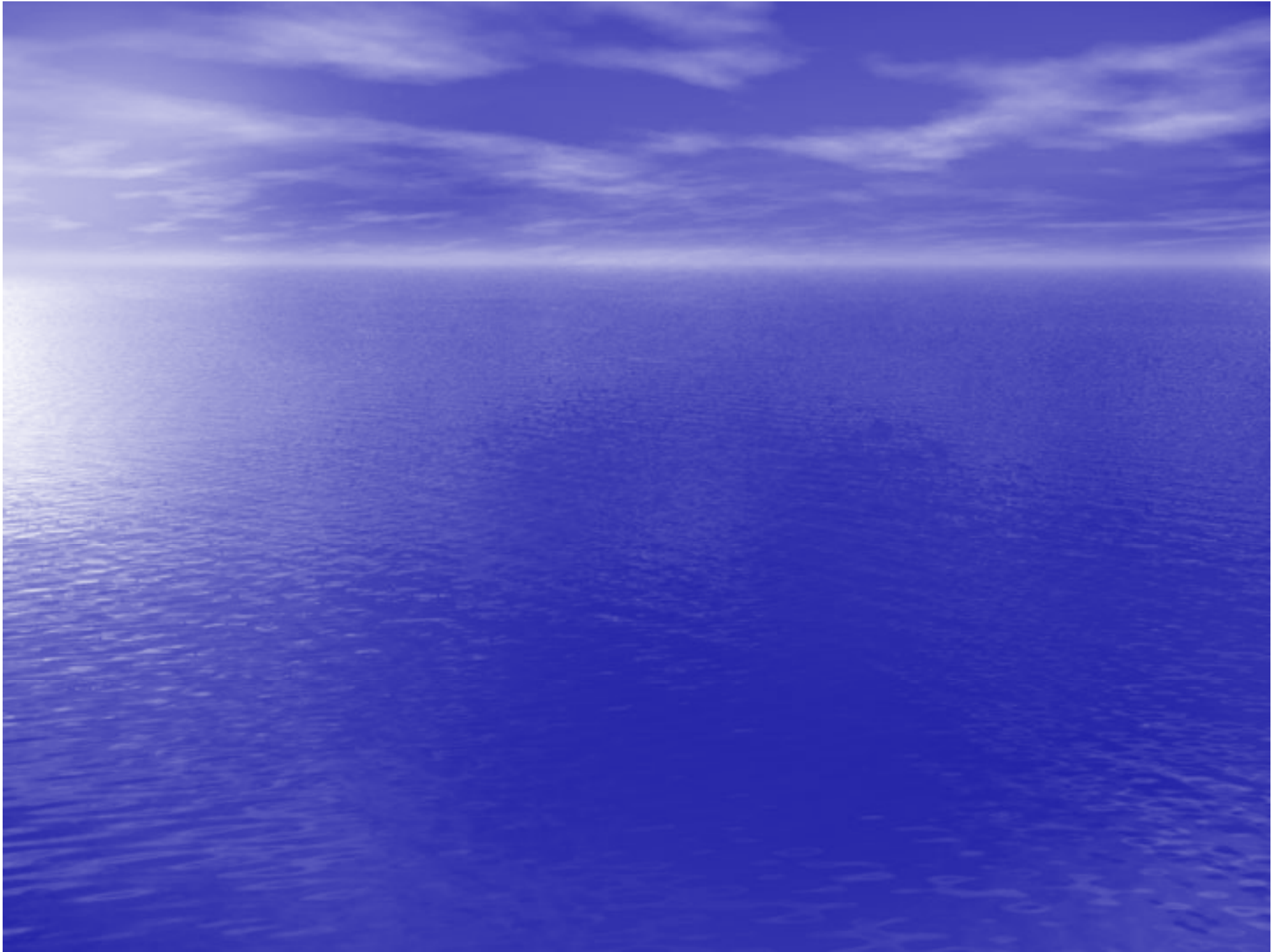


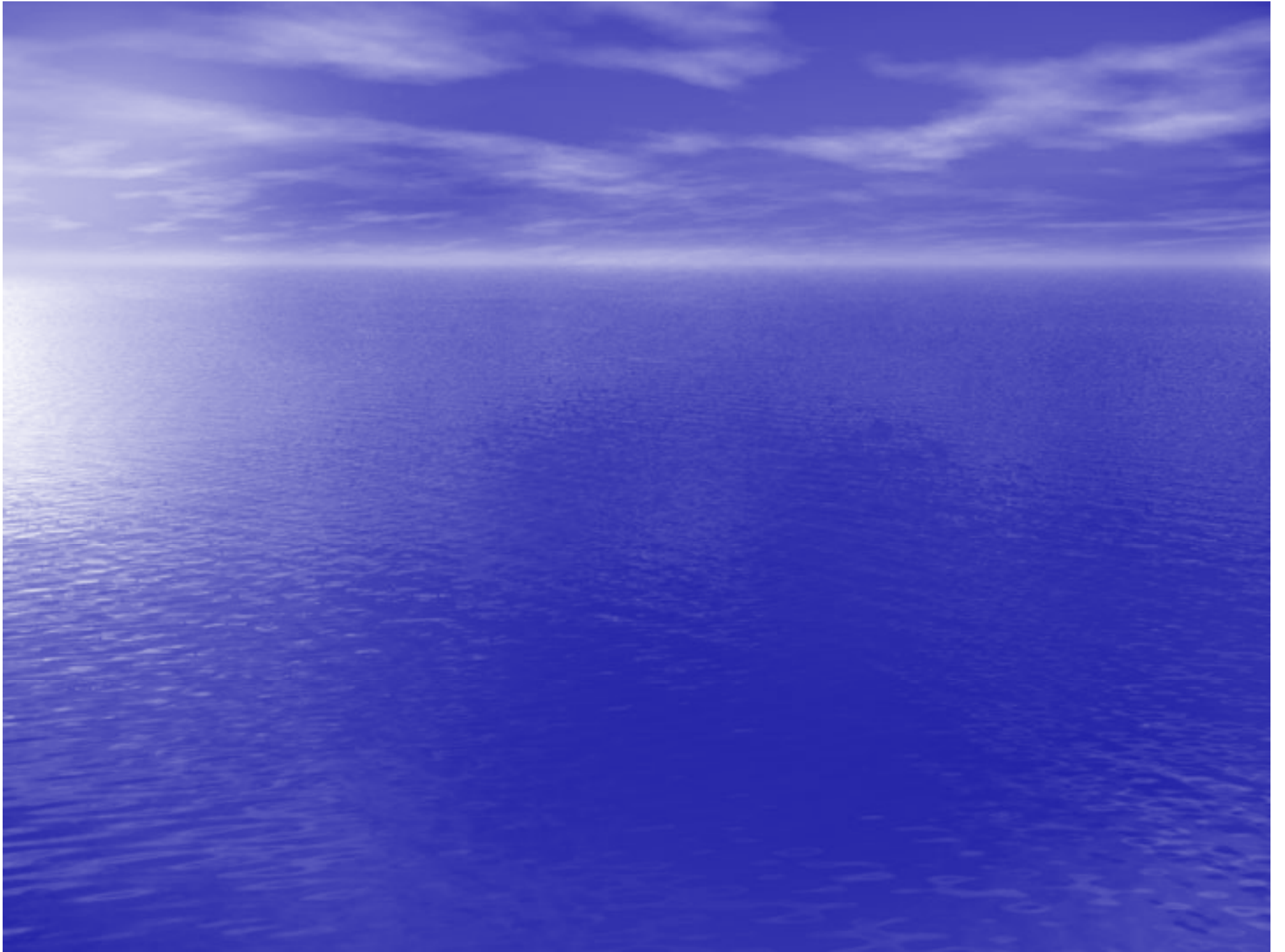
Endocrine Manifestations

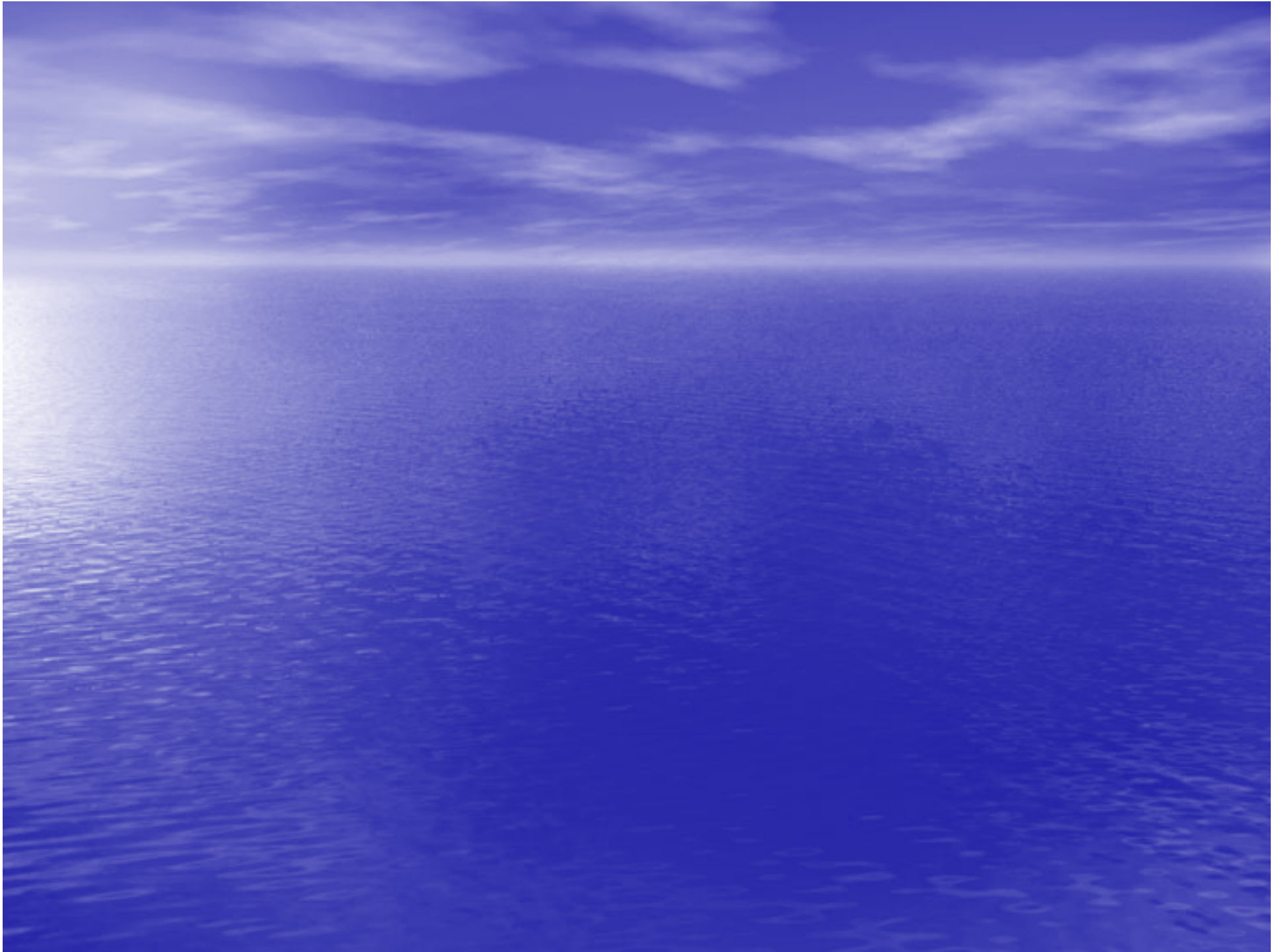
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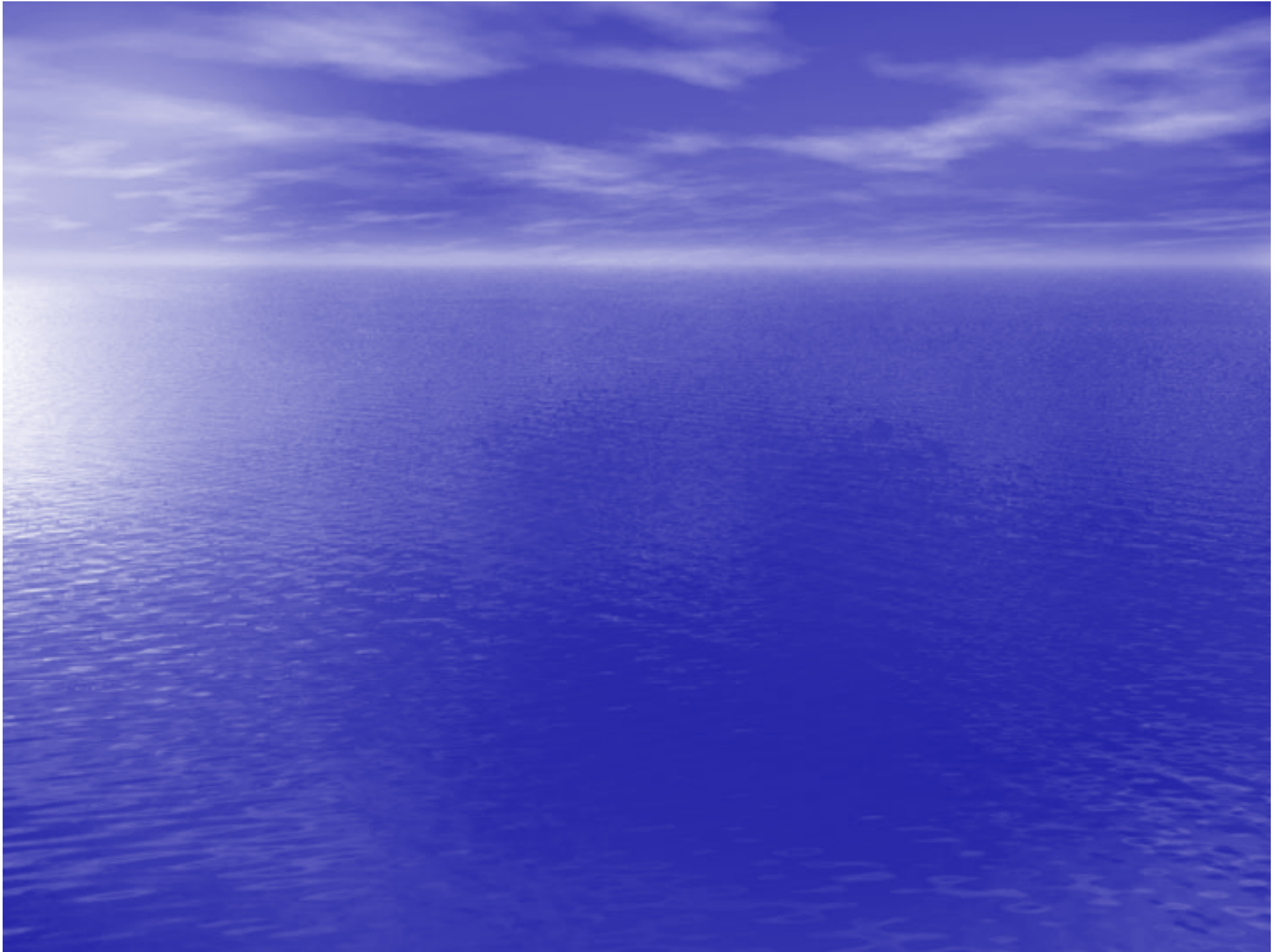


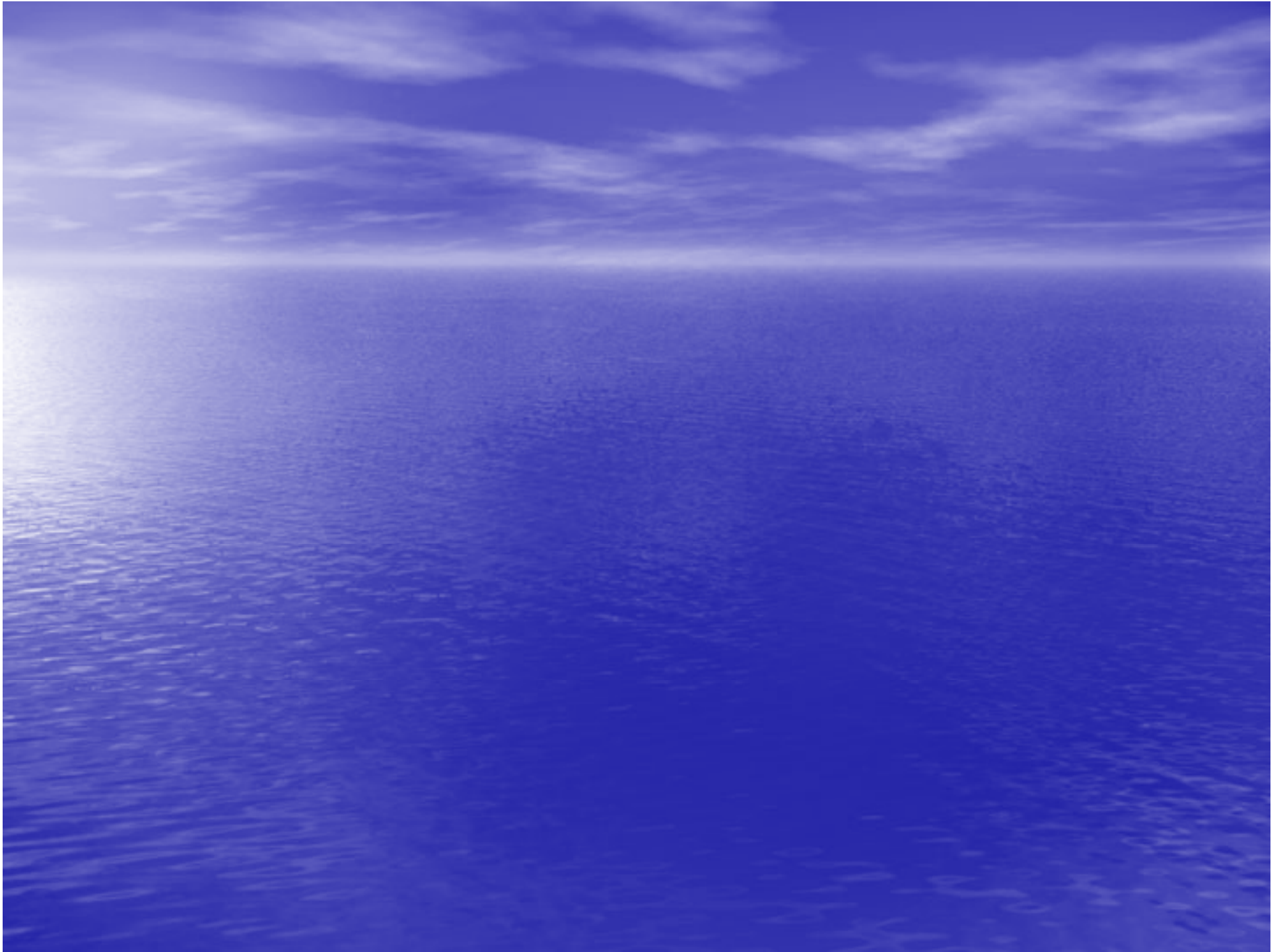






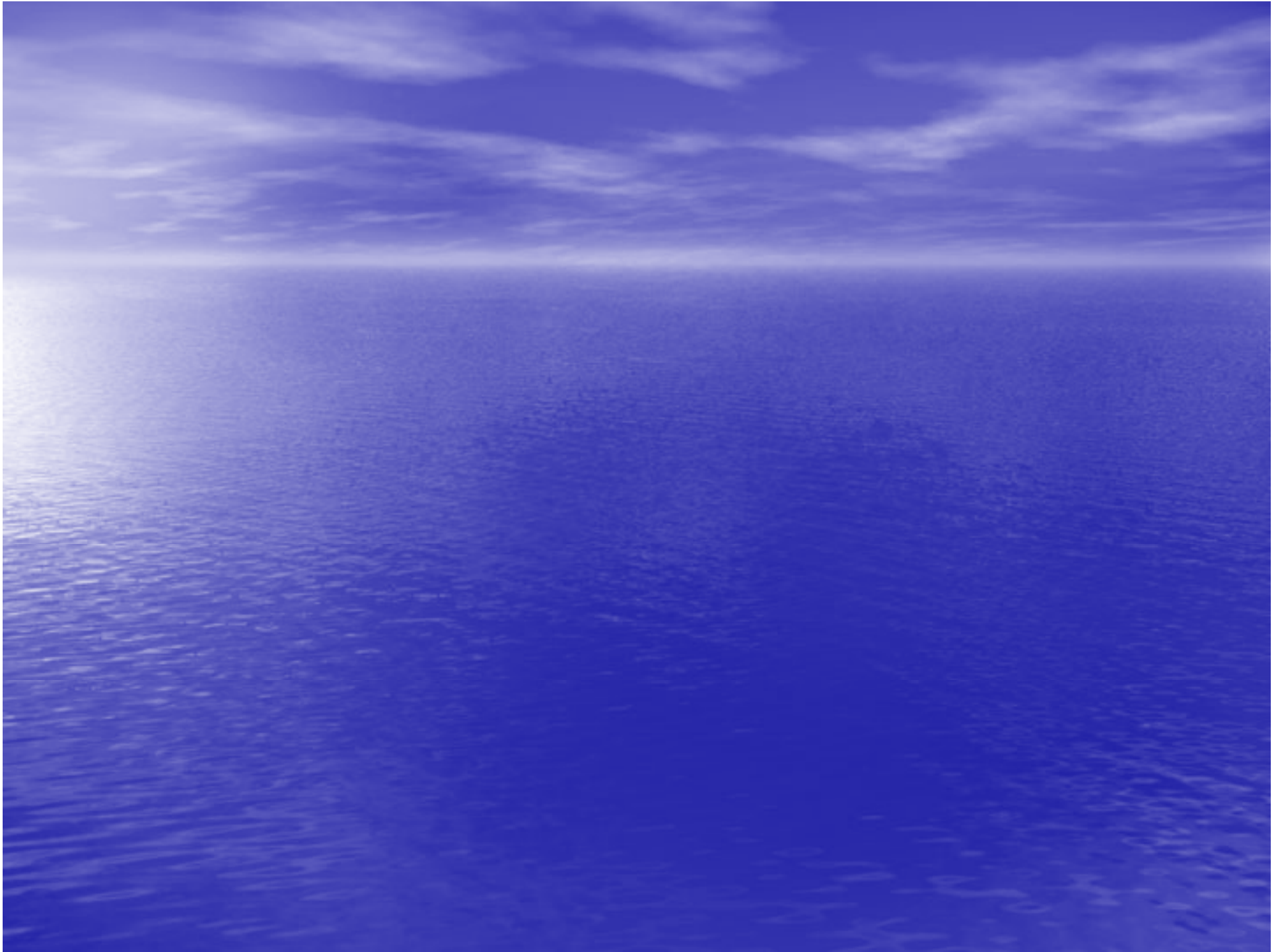


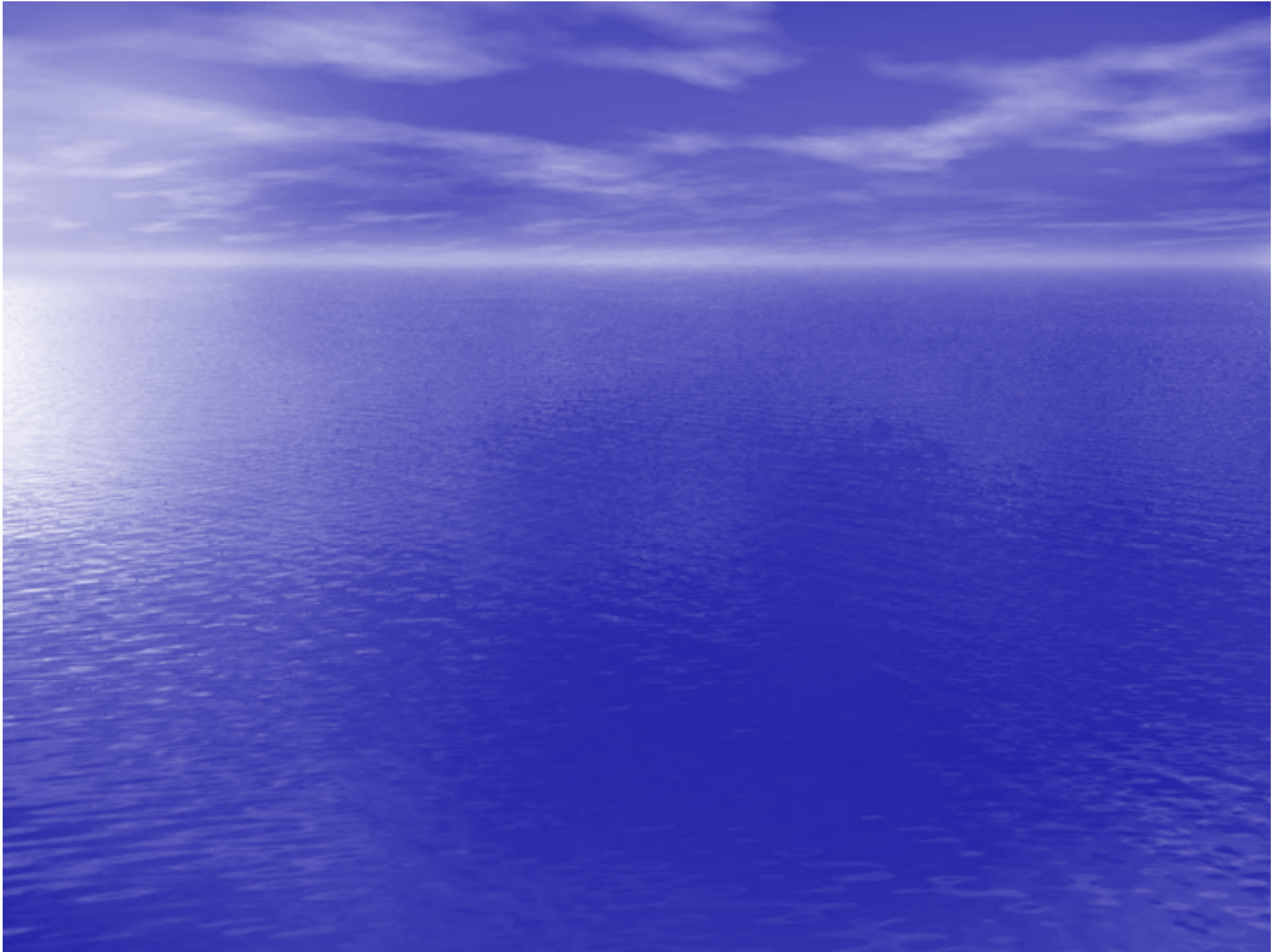


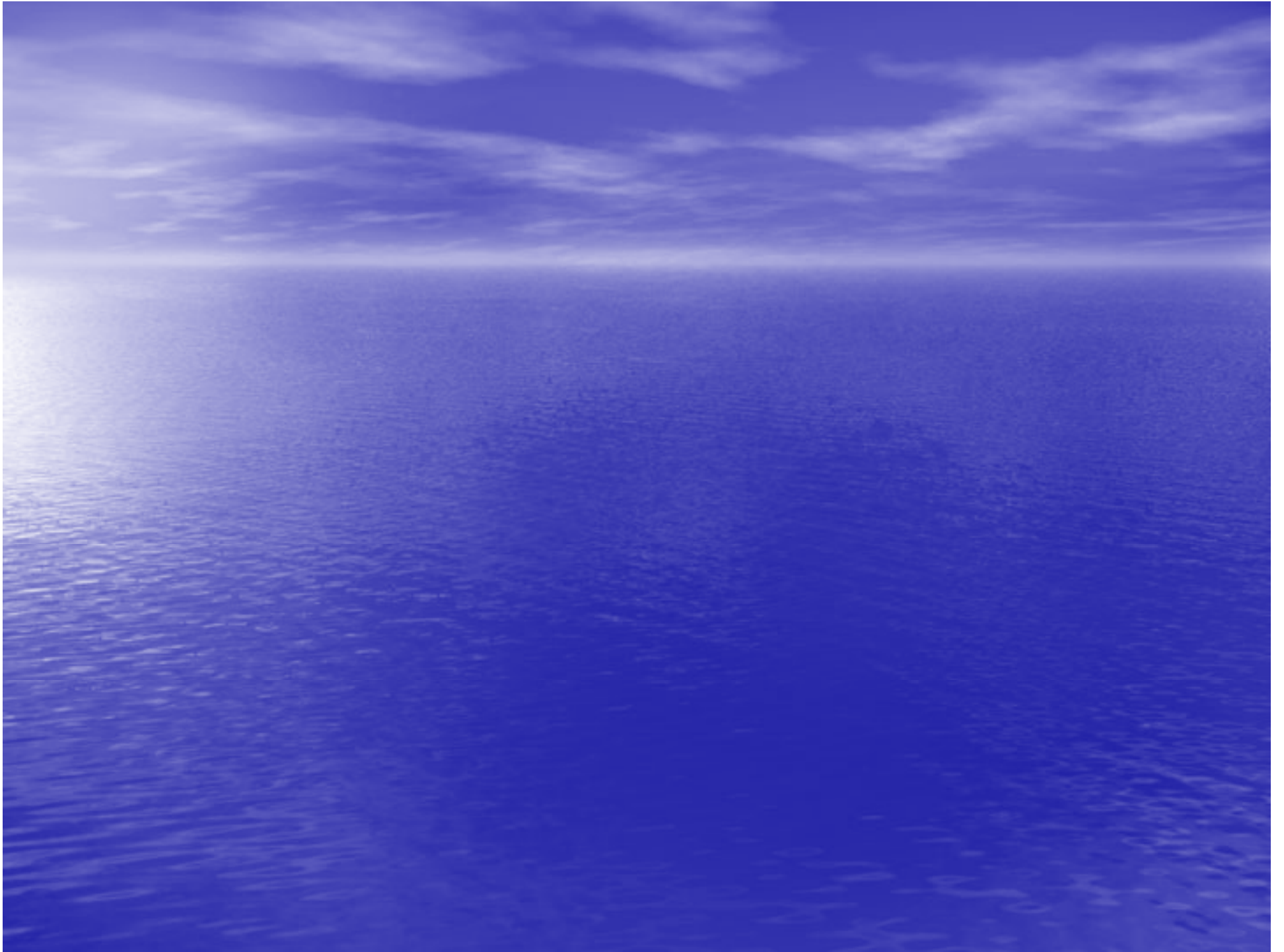


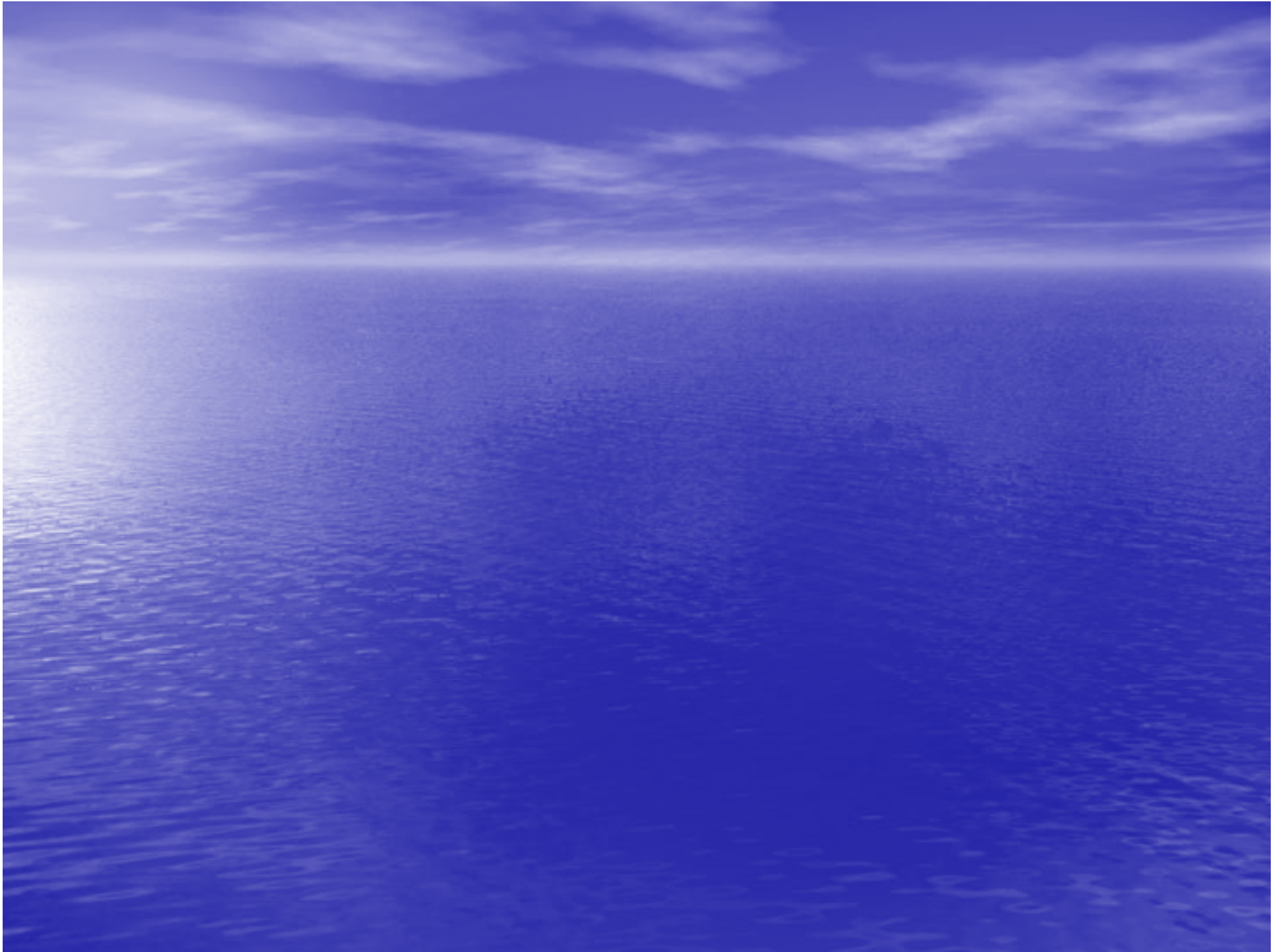
Individualized Treatment

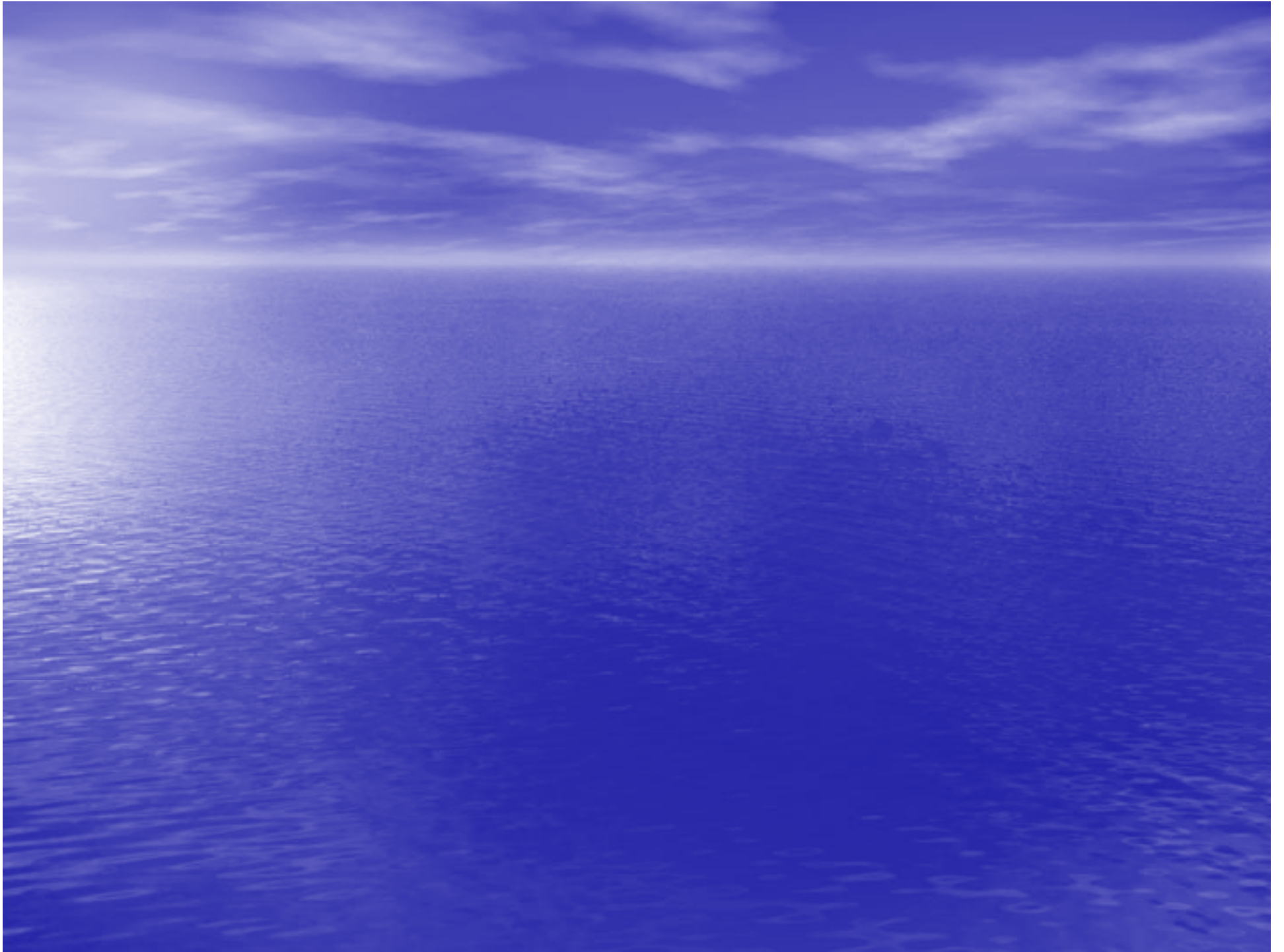
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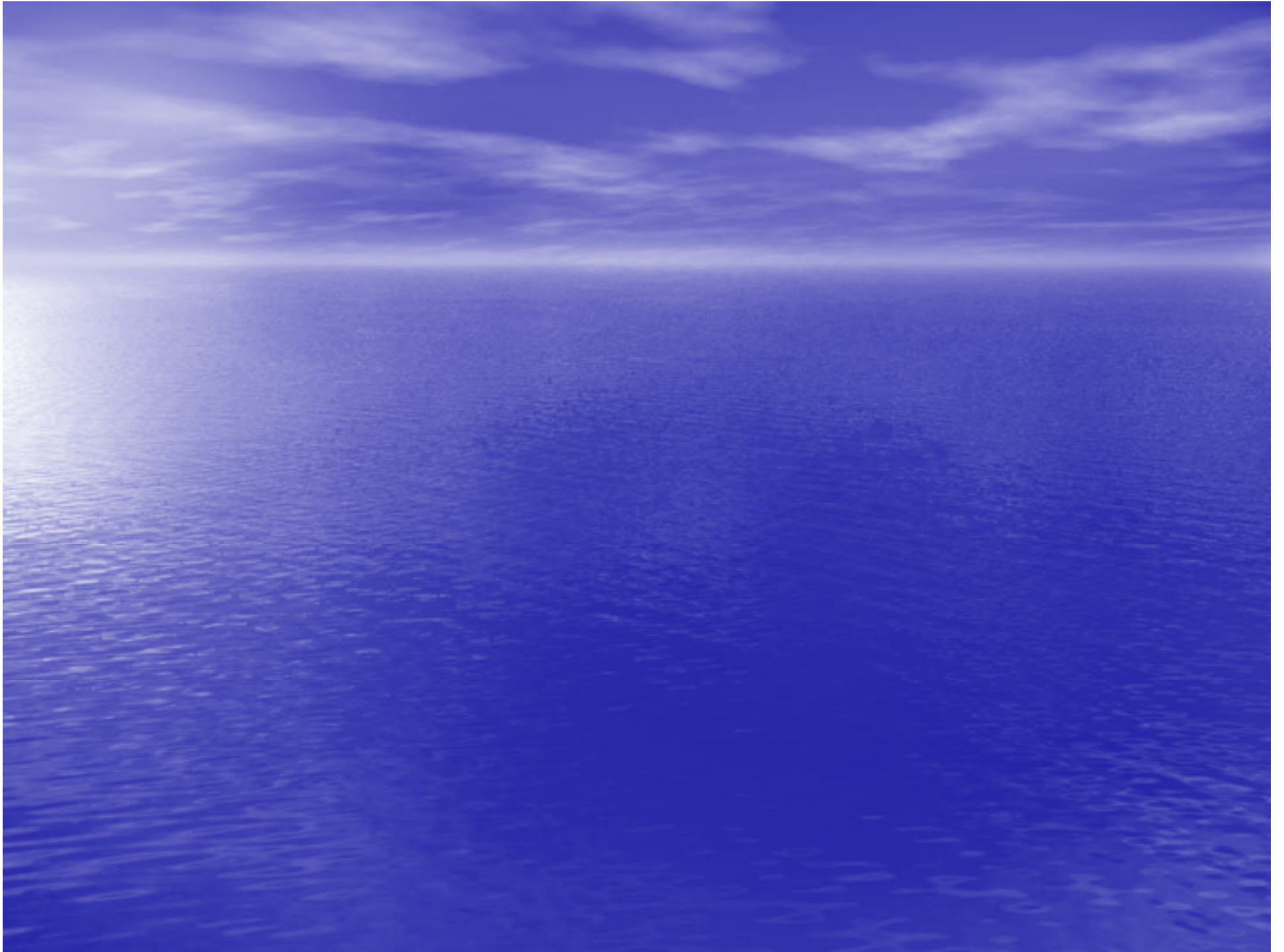






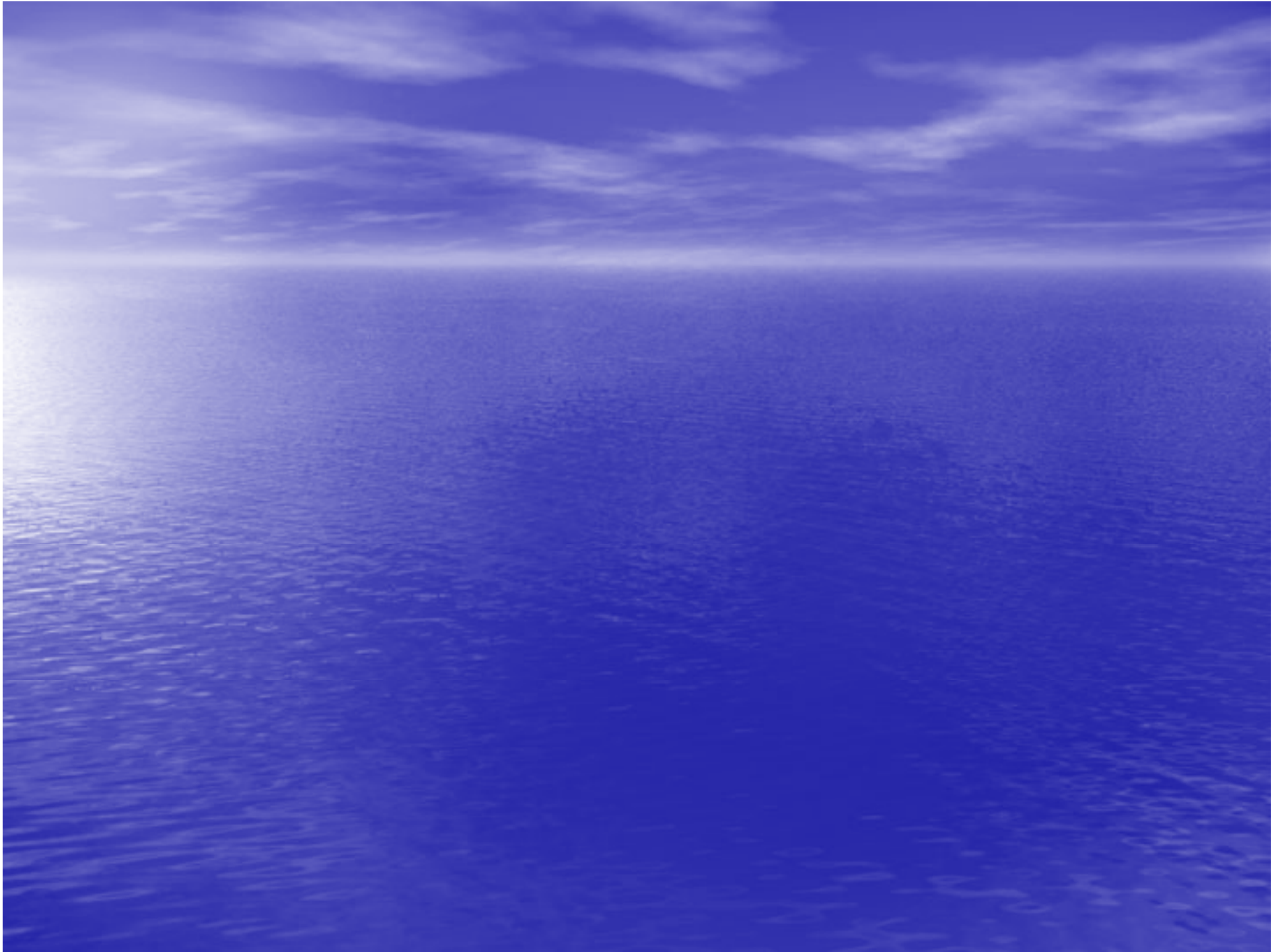


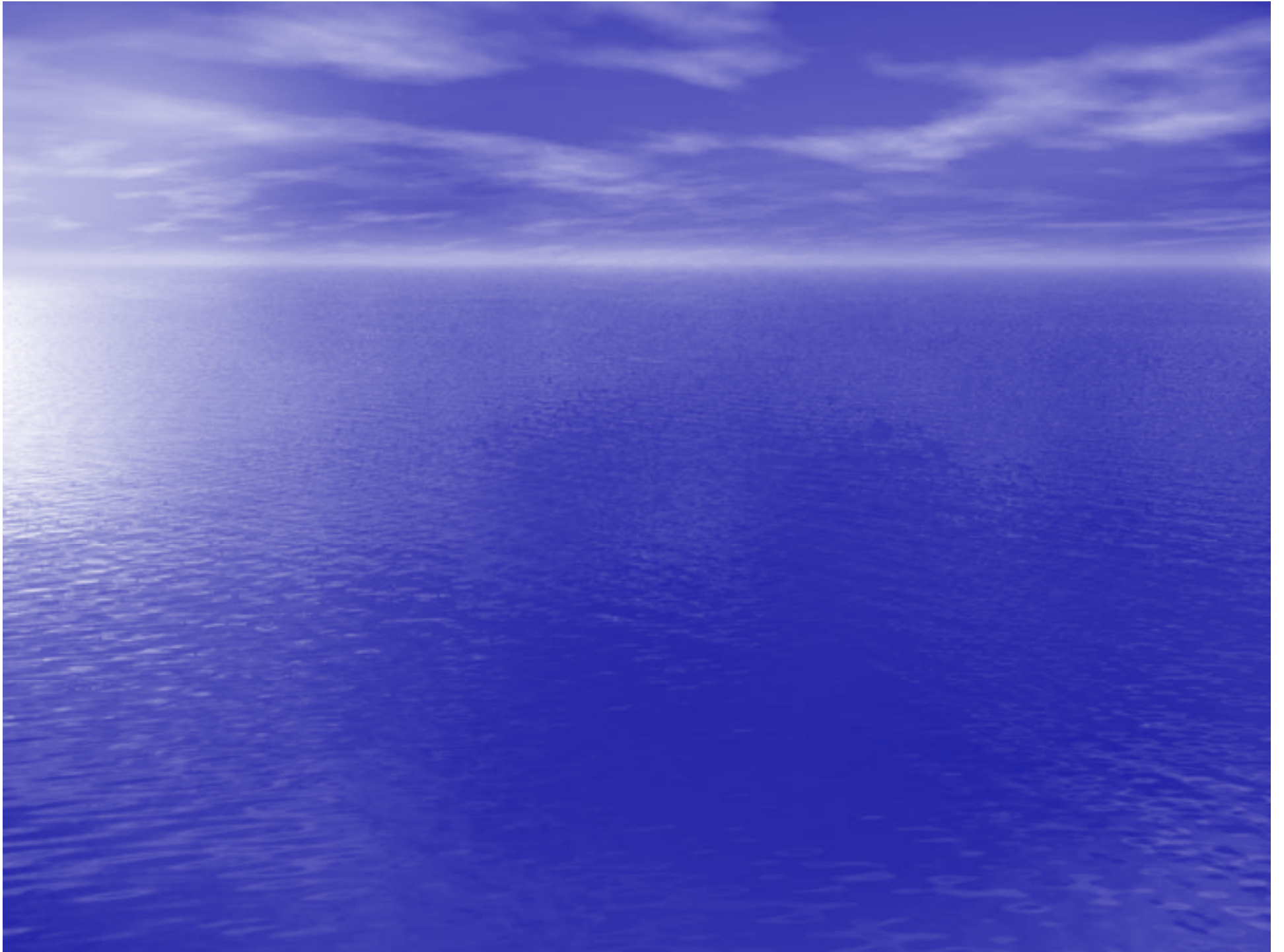


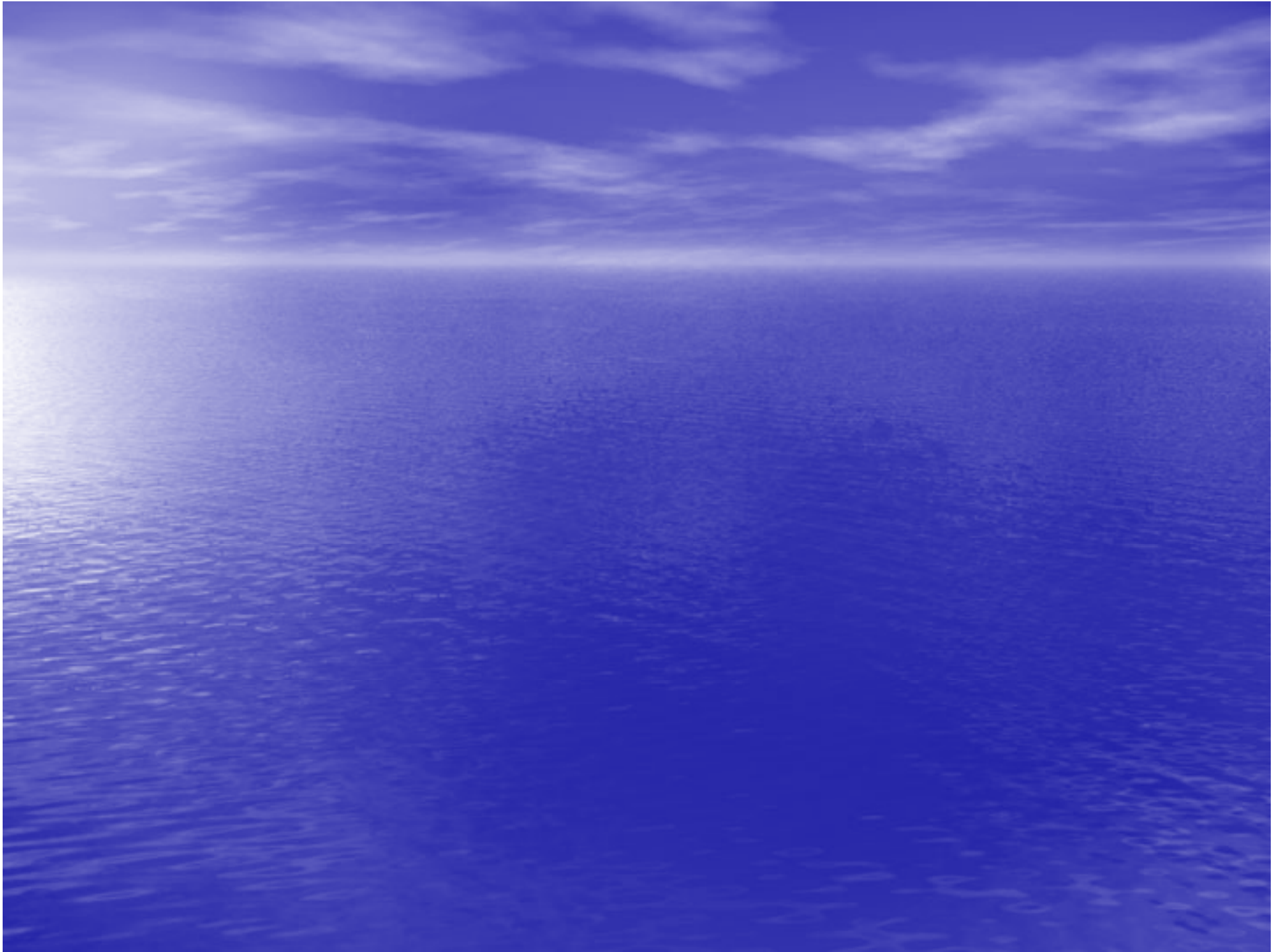


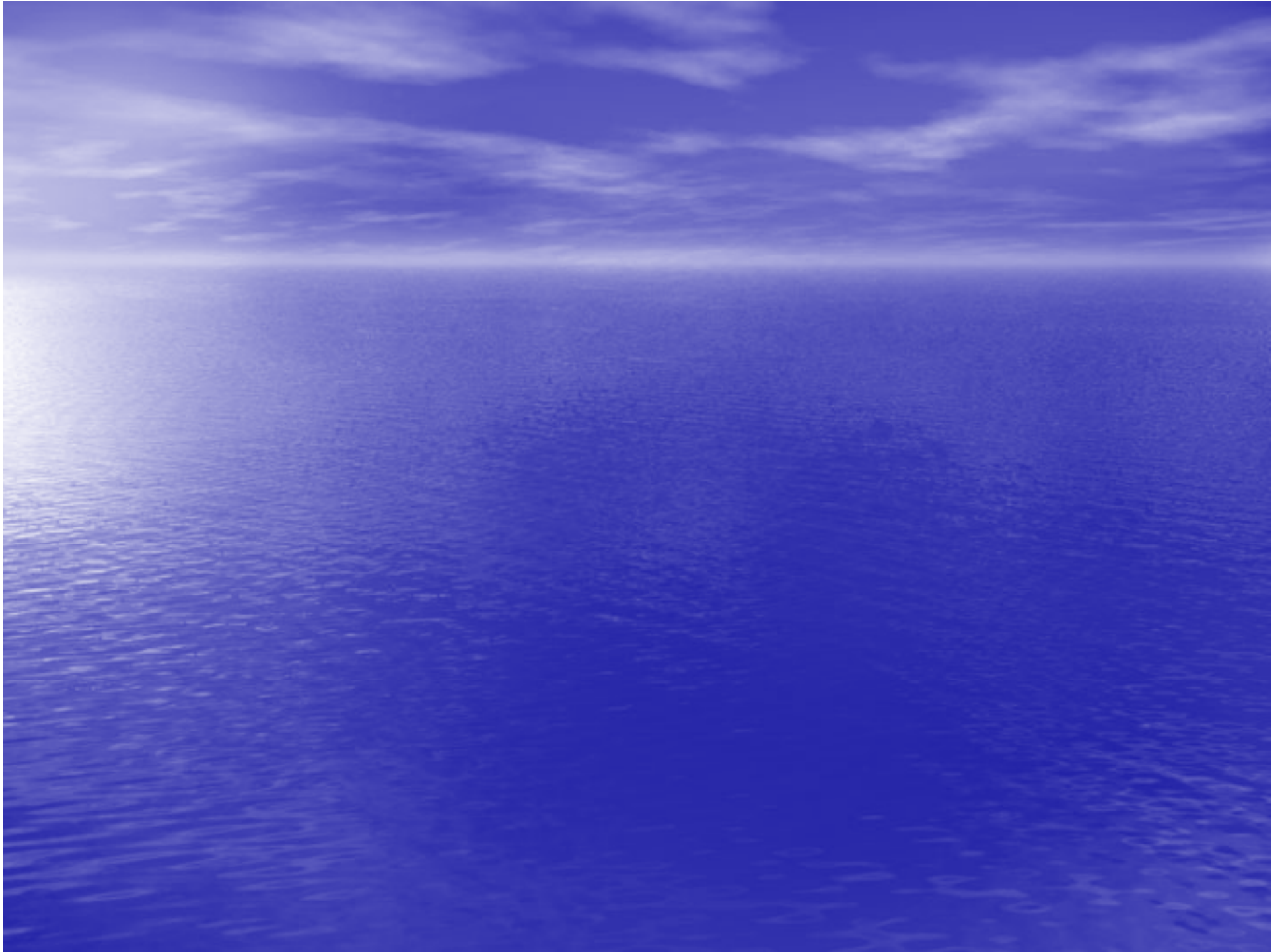
Radiation

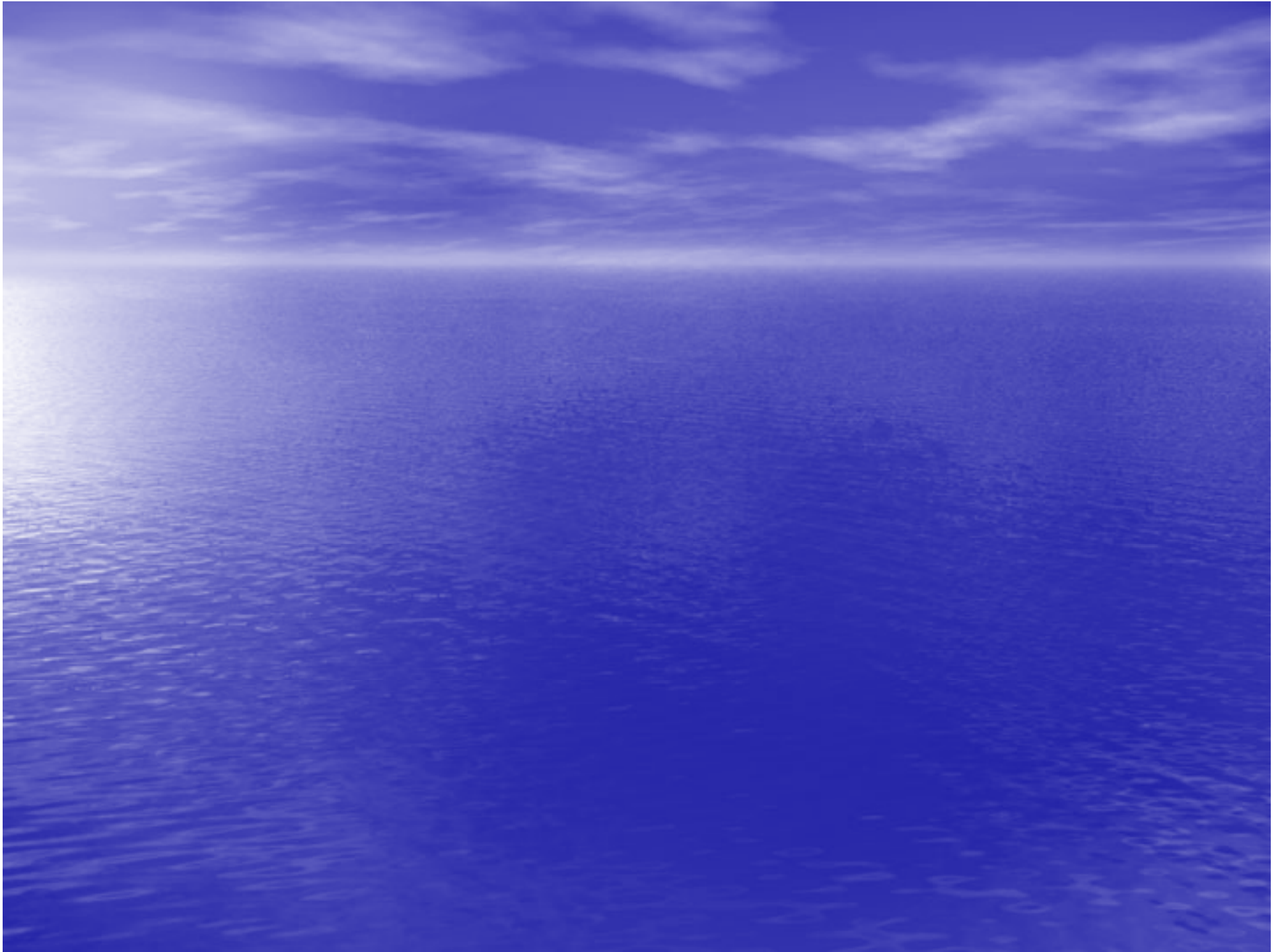
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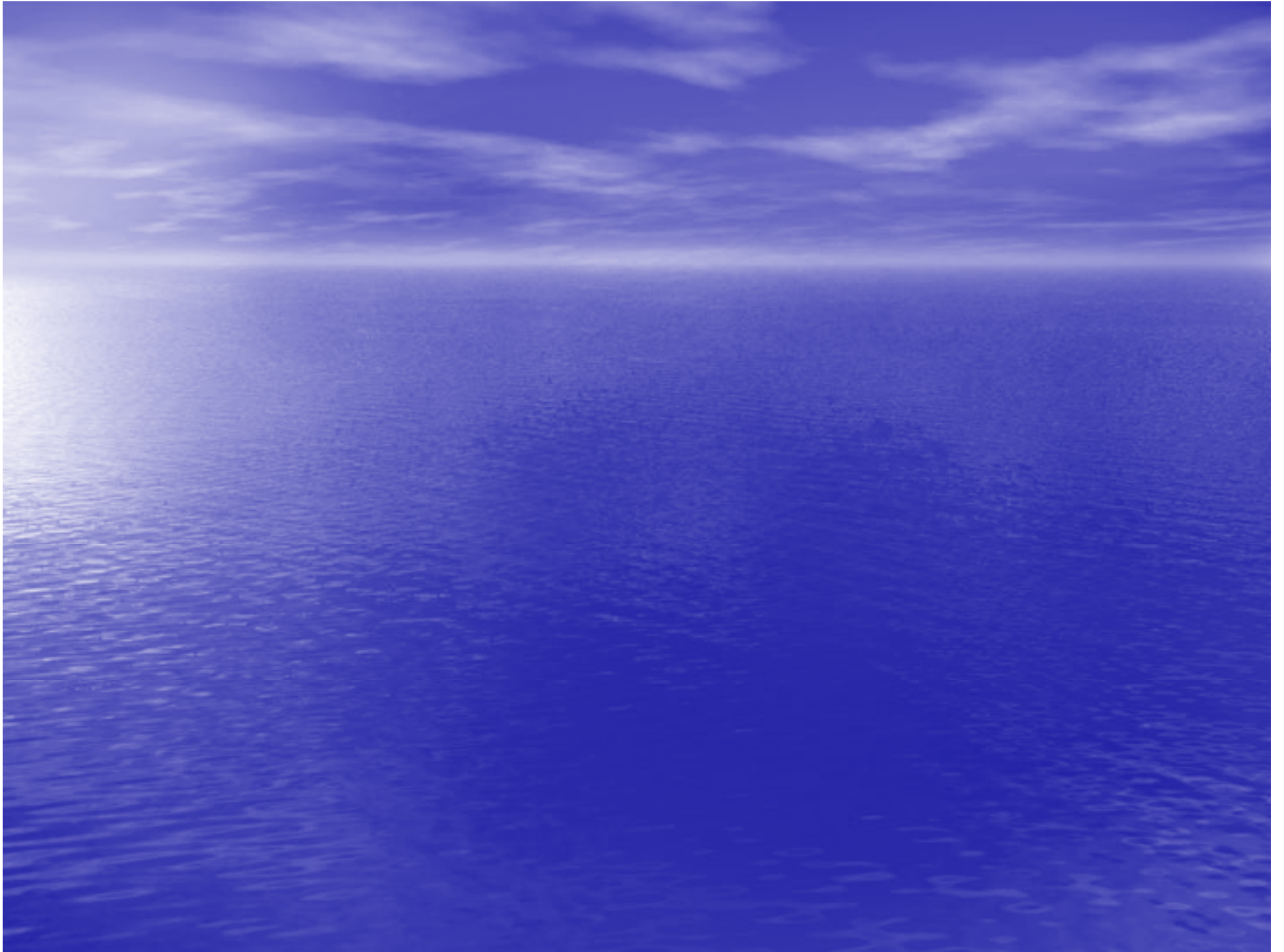












Outcomes - Literature Review

The background of the slide is a deep blue gradient. In the upper portion, there are faint, wispy white clouds. Below the clouds, a horizontal line represents a horizon. The lower two-thirds of the image are filled with a pattern of fine, concentric ripples, suggesting the surface of water. The overall effect is a calm, expansive, and slightly textured blue background.

